

# Monostotic fibrous dysplasia and solitary intramuscular myxoma of the head and neck: A unique presentation of Mazabraud's syndrome and a literature review

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**Abstract.** Mazabraud's syndrome (MS) is a rare disease that is a combination of fibrous dysplasia and intramuscular myxomas. MS is a benign lesion and there is little data on the disease due to its low incidence. In the present study, the case of a 38-year-old patient who presented with a soft-tissue mass involving the masseter and swelling at the mandibular body and mandibular ramus is reported. Since the mandible is an important aesthetic and functional organ in the oral and maxillofacial region, surgery was primarily aimed at resecting the tumor, with good safety margins, and reconstructing the resultant defect. The lesions were pathologically diagnosed as MS. The unique features of this case included the painless and monostotic fibrous dysplasia, the solitary intramuscular myxomas involving the jaw and the male gender of the patient. MS usually occurs in the lower extremities, with an unusual predilection for the right limb; however, it rarely occurs in the head and neck region. A retrospective analysis of the clinical features and management of MS was also performed in the present study, together with a literature review. From the literature, it was concluded that the incidence of MS is ~2.3-fold greater in female patients than in male patients, and that the age of onset of MS ranges between 17 and 82 years, with an average age of 46.25 years.

## Introduction

Mazabraud's syndrome (MS) is a rare benign syndrome that is reported to be associated with single or multiple intramuscular myxomas and fibrous dysplasia occurring in a single or

multiple bones. Intramuscular myxoma in itself is a relatively uncommon benign mesenchymal tumor (1), while fibrous dysplasias are more frequent benign lesions (2). To the best of our knowledge, 92 cases of MS have been reported in the medical literature thus far. However, only 9 cases have reported the combination of a solitary myxoma and monostotic fibrous dysplasia (3-10). MS involving the mandible and the adjacent soft tissue of the bone lesions was reported by Logel (11). However, MS of the masseter and the mandible are rarely reported compared with other regions. The aim of the current study was to present a novel case of solitary myxoma and monostotic fibrous dysplasia involving the jaw and the masseter. Additionally, an overview of the 92 reported cases of MS is also presented.

## Case report

On February 8th, 2014, a 38-year-old male patient presented to the Shanghai Ninth People's Hospital (Shanghai, China) with a history of a slowly growing painless swelling in the left mandibular area that had been present for 23 years. A definitive diagnosis had not been reached upon examination at a local hospital (Zhengzhou, China) in 2011. The patient had undergone a pathological biopsy prior to the current examination and was diagnosed with ossifying fibroma in another hospital (Zhongshan Hospital, Zhengzhou, China). The current physical examination identified two masses, one on the left mandible and one on the left musculus buccinator. The left mandibular lump measured ~5 cm in diameter, was hard in nature and protruded from the surface with ill-defined margins. There was no visible disease activity and no palpable tenderness. The soft-tissue swelling located in the musculus buccinator near the left mandibular measured ~3 cm in diameter; the lesion exhibited no visible ulceration and no palpable tenderness (Fig. 1). Conventional panoramic radiography demonstrated a hazy shadow of low but uneven density, often with ill-defined margins in the mandible. The patient did not present with any bony deformities or café-au-lait spots. Computed tomography (CT) and three-dimensional CT reconstruction in the oral and maxillofacial regions showed that the mass exhibited a localized 'ground-glass' pattern on the left mandibular body and mandibular ramus, and an uneven high-density shadow

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**Key words:** Mazabraud's syndrome, fibrous dysplasia, intramuscular myxomas, mandible

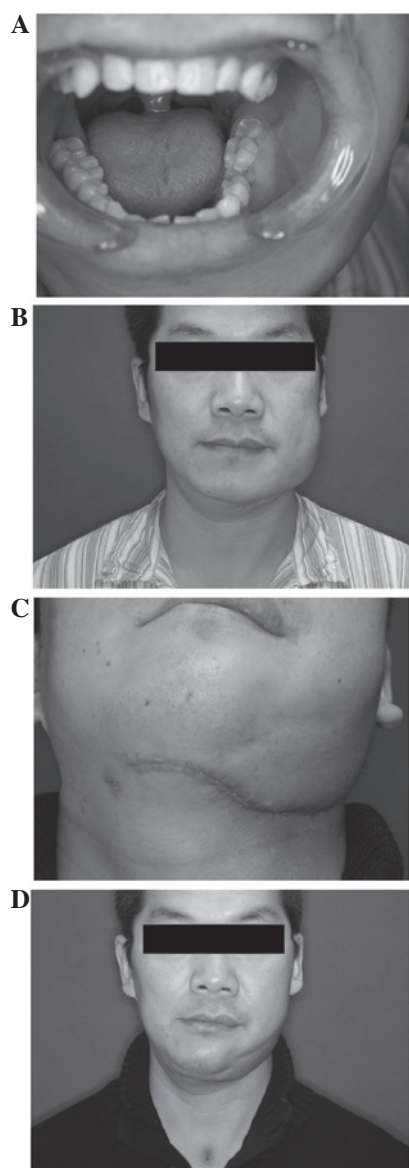


Figure 1. (A) Pre-operative intraoral view of the mass located in the left jaw. (B and C) Frontal view of the patient at 1 week post-surgery demonstrating swelling of the left side of the jaw. (D) Frontal view showing no marked contour defect of the jaw at 3 months post-surgery.

was identified (Fig. 2). The buccal bone cortex of the mass was discontinuous. Adjacent soft tissue was markedly thickened, with uniform density.

A pre-operative diagnosis of single intramuscular myxoma and a benign bone lesion was determined and histopathologically confirmed following resection of the lesions. The left mandible lesions and outermost region of the bony plates of the ramus were completely resected, while conserving the condyle. Next, a free fibular osteocutaneous flap with skin island reconstruction was performed. During the surgery, a soft-tissue mass of 3.5x2.5x1.0 cm was resected. Histological examination of the left mandibular lesions showed immature woven bone trabeculae devoid of osteoblastic lining laid in proliferative fibrous connective tissue, and the lesions were characterized by some mucous degeneration and were hypercellular enough to be characterized as fibrous dysplasia. The histopathological examination of the buccal mass revealed a tumor extending

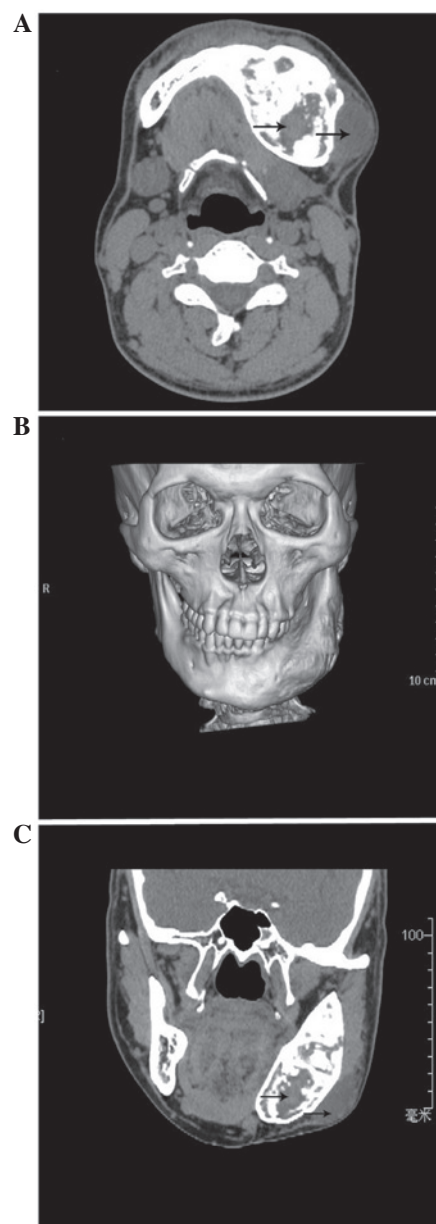


Figure 2. (A) Coronal computed tomography (CT) in the oral and maxillofacial regions showing a solid soft-tissue mass within the left musculus buccinator (right arrow) and radiolucent cystic lesions with frosted glass margins in the left body of the mandible (left arrow) (B) Three dimensional CT reconstruction in the oral and maxillofacial regions showing bone widening and local destruction of the adjacent cortex. (C) Axial CT in the oral and maxillofacial regions showing a solid soft tissue mass within the left musculus buccinator (right arrow) and radiolucent cystic lesions with frosted glass margins in the left body of the mandible (left arrow).

into the striated muscle, formed from hypocellular tissue with bland stellate cells and spindle-shaped cells within a polysaccharide abundant myxoid matrix. A histopathological diagnosis of intramuscular myxoma was formed. Immunohistochemical analysis showed positive staining for vimentin and cluster of differentiation (CD)99, and negative staining for desmin, smooth muscle actin, B-cell lymphoma-2, S-100 protein and CD34. The intramuscular myxoma with adjacent fibrous dysplasia was diagnosed as MS (Fig. 3). Follow-up examinations were planned every 3-6 months for 3 years. Three and six months after surgery, MRI follow-up examinations revealed that the

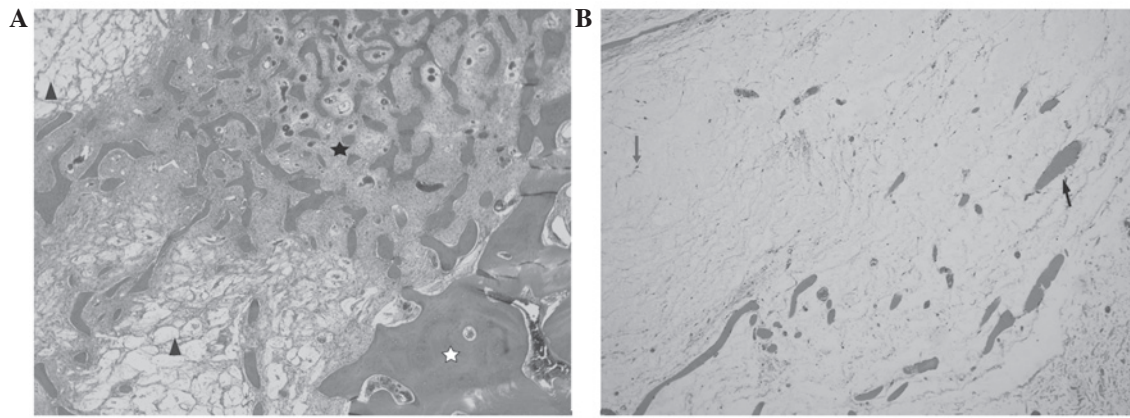


Figure 3. (A) Histological specimen showing fibrous dysplasia composed of immature woven bone trabeculae devoid of osteoblastic lining (black star) and the lesions of segmental myxoid (black triangle) and normal bone (white star) tissues (hematoxylin and eosin; original magnification, x40). (B) Histological specimen showing the myxoma (grey arrow) with a well-defined border and the adjacent skeletal muscle (black arrow) (hematoxylin and eosin; original magnification, x100).

patient exhibited no clear evidence of recurrence of myxoma and fibrous dysplasia. However, the patient has subsequently been lost to follow-up.

This study was approved by the Shanghai Ninth People's Hospital Institutional Review Board and the patient provided written informed consent.

## Discussion

MS was first described in the German literature by Henschen in 1926 (12), and Mazabraud *et al* presented a pattern of association between fibrous dysplasia and soft-tissue myxomas in 1967 (13). Only a limited number of cases appear in the literature. Not including the present case, ~92 cases of what has come to be termed as 'Mazabraud's syndrome' have been described in the literature to date (Table I) (3,5-75).

MS is an uncommon type of benign tumor that mainly occurs in the lower limbs (14). In the most extensive review of MS to date in the literature, the present study found 9 cases of the unusual association of monostotic fibrous dysplasia combined with a rare solitary myxoma. A mandibular origin accounted for ~10 cases.

Six of these cases were excluded from the study because we were not get detailed information (3,6,9,15-17), therefore, including the present case, a total of 87 cases were reviewed. Table I reveals that the majority of affected patients were female (61 cases). While only 26 cases were male, indicating an ~2.3-fold greater incidence in females than in males. The most common onset of fibrous dysplasia is polyostotic, present in 70 cases (80.46%). In 17 cases, fibrous dysplasia was monostotic (19.54%). However, these myxomas were solitary in 40 cases (45.98%). Monostotic fibrous dysplasia and solitary myxoma, when associated, is a rare condition. Only a few cases were mentioned in the literature. Another common finding of MS that was recorded in the literature was its localization in the lower extremities, which occurred in 82.71% of cases (67 in the lower extremities, 35 in the upper extremities, 19 in all four extremities and in 11 in the head and neck; in 6 cases, detailed information was not recorded). These tumors were bilateral in 30 cases and unilateral in 51 cases. This finding

is in agreement with other studies, which confirm that the majority patients with MS have multiple intramuscular myxomas and polyostotic fibrous dysplasia that tend to be located in the lower extremities, with a particular predilection for the lower right limb (18). The age of onset of MS in the 87 patients studied ranged between 17 and 82 years, with an average age of 46.25 years. MS was most commonly diagnosed in the middle-aged population.

The case reported in the present study differs from the majority of the previously reported cases due to its rare localization in the head and neck, specifically the mandible, in addition to the unusual association of monostotic fibrous dysplasia and solitary myxoma. As the mandible is an important aesthetic and functional organ in the oral and maxillofacial region, an accurate pre-operative estimation of mandibular invasion remains challenging.

Another unusual aspect of the present study was the complex appearance of histopathological findings of the mucous degeneration of fibrous dysplasia. It is rare to find the mucous transformation of fibrous dysplasia that was observed in this case in patients with MS. Although uncommon, malignant transformation of fibrous dysplasia can also occur in patients with MS (76).

Intramuscular myxomas are rare benign tumors of the musculoskeletal system that were first recognized as a definite clinicopathologic entity by Enzinger in 1965. The study presented a brief compilation of 34 examples of intramuscular myxoma and demonstrated the benign clinical course of the tumor, as relatively few of the cases exhibited a tendency to metastasize (77). While intramuscular myxoma is rare, fibrous dysplasia is not. Fibrous dysplasia is a congenital disease characterized by a condition affecting one, several or numerous bones, leading to osteolytic lesions, deformities and fractures (78). Intramuscular myxoma is usually a solitary lesion, and exhibits an association with multiple fibrous dysplasia (19). Occasionally, MS is associated with McCune-Albright syndrome (MAS) (20). MAS is a rare syndrome with the three combined characteristics of polyostotic fibrous dysplasia, endocrine dysfunction and café-au-lait spots (79).

Table I. Reported cases of Mazabraud's syndrome.

First author/s, year (ref.)	Gender	Age, years	Details of lesion		Lesion localization
			Fibrous dysplasia	Myxoma	
Henschen, 1926 (12)	Female	66	Monostotic	Multiple	R-L
Krogus, 1929 (23)	Female	26	Polyostotic	Multiple	B-U/B-L
Uehlinger, 1940 (24)	Male	67	Polyostotic	Single	R-L
Braunchwarth, 1953 (5)	Female	55	Polyostotic	Single	R-L
Mazabraud and Girard, 1957 (25)	Male	54	Polyostotic	Multiple	R-U'R-L
Heinemann and Woerth, 1958 (26)	Female	82	Polyostotic	Multiple	
Laporte, 1961 (27)	Female	24	Polyostotic	Single	
Lick, 1962 (28)	Male	59	Polyostotic	Multiple	
Mazabraud, 1967 (13)	Female		Polyostotic	Multiple	
Roze, 1967 (29)	Female	38	Polyostotic	Multiple	B-U/B-L
Semat, 1969 (30)	Male	47	Polyostotic	Single	B-U/B-L
Wirth <i>et al</i> , 1971 (31)	Male	17	Polyostotic	Multiple	B-U/B-L
	Male	33	Polyostotic	Multiple	B-U/B-L'H&N
Lejeune, 1972 (32)	Male	41	Polyostotic	Multiple	B-U/B-L
Ireland, 1973 (33)	Female	49	Monostotic	Multiple	B-U/B-L
	Female	59	Polyostotic	Multiple	R-L
	Female	28	Polyostotic	Multiple	R-L
Logel, 1976 (11)	Female	41	Polyostotic	Multiple	B-U/B-L'H&N
Berkhoff, 1981 (6)			Monostotic	Single	
Sedmak, 1983 (34)	Male	50	Polyostotic	Single	L-L
Lever and Pettingale, 1983 (35)	Female	50	Polyostotic	Single	B-U/B-L
Segev and Reiner, 1985 (3)					
Witkin, 1986 (36)	Male	40	Polyostotic	Single	B-L
Blasier, 1986 (37)	Female	57	Polyostotic	Multiple	L-U/L-L
Biagini, 1987 (38)	Female	42	Polyostotic	Multiple	B-U/B-L
Glass-Royal, 1989 (39)	Male	42	Polyostotic	Single	B-U'H&N
Sundaram, 1989 (40)	Female	31	Polyostotic	Multiple	R-L
Gianoutsos, 1990 (41)	Male	53	Monostotic	Multiple	R-L
Prayson, 1992 (42)	Female	36	Polyostotic	Single	R-U
Gober and Nicholas, 1993 (43)	Female	37	Monostotic	Multiple	L-L
Aoki, 1995 (44)	Female	67	Polyostotic	Multiple	B-U/B-L
	Female	46	Polyostotic	Single	R-L
Fujii, 1996 (45)	Female	47	Monostotic	Single	B-U/B-L
Limouzy, 1996 (7)	Male	45	Monostotic	Single	R-L
Court-Payen, 1997 (46)	Female	50	Polyostotic	Multiple	R-U'H&N
Szendroi, 1998 (22)	Female	52	Polyostotic	Multiple	B-L
	Female	34	Polyostotic	Multiple	B-L
	Female	54	Polyostotic	Single	L-L
Cabral, 1998 (47)	Male	44	Polyostotic	Multiple	R-L'H&N
Lopez-Ben, 1999 (21)	Female	40	Polyostotic	Multiple	B-U/B-L'H&N
Thomacot, 1999 (48)	Female	36	Polyostotic	Single	L-L
Walker, 1999 (49)	Male	36	Polyostotic	Single	R-L
Kransdorf and Murphey, 1999 (50)	Female	67	Monostotic	Multiple	R-L
Okamoto, 2000 (51)	Female	53	Polyostotic	Multiple	
	Female	34	Polyostotic	Multiple	B-U/B-L
	Female	38	Polyostotic	Single	L-L
Struk, 2000 (8)	Female	53	Monostotic	Single	R-L
Faivre, 2001 (52)	Female	35	Polyostotic	Multiple	B-U/B-L
	Male	42	Polyostotic	Multiple	L-U'R-L
Delabrousse, 2001 (53)	Female	40	Monostotic	Multiple	R-L
Pollandt, 2002 (54)	Female	42	Polyostotic	Multiple	B-L



Table I. Continued.

First author/s, year (ref.)	Gender	Age, years	Details of lesion		Lesion localization
			Fibrous dysplasia	Myxoma	
Iwasko, 2002 (55)	Female	39	Polyostotic	Multiple	L-L
	Female	39	Polyostotic	Multiple	B-U/B-L'H&N
	Female	63	Polyostotic	Multiple	B-L
	Female	52	Monostotic	Multiple	R-U'R-L
	Male	49	Polyostotic	Single	L-L
	Female	65	Polyostotic	Single	
	Female	56	Polyostotic	Single	R-U
Tsitouridis, 2002 (56)	Female	28	Polyostotic	Single	B-U/B-L'H&N
Fang, 2003 (57)	Male	33	Polyostotic	Single	R-L
Jhala, 2003 (58)	Female	44	Polyostotic	Multiple	R-U
Kabukcuoglu, 2004 (19)	Female	52	Polyostotic	Single	L-L
Nguyen and Ram, 2005 (59)	Male	48	Polyostotic	Multiple	R-U
Endo, 2007 (75)	Male	64	Monostotic	Single	R-L
	Female	64	Monostotic	Single	R-L
Miyake, 2006 (9)					
Martin, 2007 (60)	Female	52	Polyostotic	Multiple	L-L
McLaughlin, 2007 (61)	Female	53	Polyostotic	Single	L-U
Singnurkar, 2007 (62)	Male	42	Polyostotic	Multiple	H&N
Calisir, 2007 (63)	Female	65	Monostotic	Multiple	L-L
MacFarlane, 2007 (64)	Male	45	Polyostotic	Multiple	B-U/B-L
Schepers, 2008 (65)	Male	35	Polyostotic	Single	L-U
Santos, 2008 (66)	Male	48	Polyostotic	Single	H&N
Zhao, 2008 (10)	Female	60	Monostotic	Single	R-L
Beele, 2008 (15)					
Tagliafico, 2009 (67)	Female	40	Polyostotic	Single	L-L
Crawford, 2009 (12)	Male	63	Polyostotic	Single	R-U
Tang, 2009 (16)					
Zoccali, 2009 (14)	Female	32	Polyostotic	Single	R-L
Arishima, 2010 (3)	Female	71	Monostotic	Single	L-L
Case, 2010 (68)	Female	69	Polyostotic	Single	L-U
Ijpma, 2010 (69)	Female	51	Polyostotic	Single	B-L
Kitagawa, 2011 (20)	Female	49	Polyostotic	Multiple	B-U
van der Wal, 2011 (70)	Female	49	Polyostotic	Single	L-L
Yang, 2011 (17)					
Gaumétou, 2012 (71)	Male	56	Polyostotic	Multiple	B-L
Schimmöller, 2012 (72)	Female	40	Monostotic	Multiple	L-U
John, 2013 (73)	Female	25	Polyostotic	Multiple	B-U/B-L
	Female	25	Polyostotic	Single	L-L
Munksgaard, 2013 (18)	Female	54	Polyostotic	Multiple	B-L
Tsourdi, 2013 (74)	Female	45	Polyostotic	Single	B-L
Present study	Male	38	Monostotic	Single	H&N

R-L, right lower limbs; B-U/B-L, bilateral upper limbs/bilateral lower limbs; L-L, left lower limbs; L-U, left upper limbs; R-U, right upper limbs; H&N, head and neck.

The clinical presentation of MS is similar to that of fibrous dysplasia and intramuscular myxoma. Myxoma may be diagnosed in adolescents or young adults with symptoms, but fibrous dysplasia is incidentally found on imaging studies. In a series of 66 patients, Zoccali *et al* (14) reported that the myxoma develops

12 years prior to the fibrous dysplasia diagnosis. Intramuscular myxoma is a rare, benign tumor of the musculoskeletal system, with a low incidence. The tumor may occur in isolation or in association with fibrous dysplasia or McCune-Albright syndrome (21). Pain and facial palsy are uncommon. The tumor

can occur at any location and tends to involve the large muscles of the thighs, followed by the buttocks, arms, and chest wall and shoulders. The majority of intramuscular myxomas are two or more, painless, palpable masses that are firm, slightly movable and discrete. Moreover, the myxomas are most typically located in the area of the bone lesions (21). Fibrous dysplasia of the bone is a disease that is characterized by bone deformities, pain and pathological fractures. Patients can, however, be asymptomatic. Fibrous dysplasia can involve one bone (monostotic) or several bones (polyostotic) (80). There are few conditions which present as a solitary intramuscular mass in association with monostotic fibrous dysplasia. MS occurs at a younger age, may be associated with additional symptoms (café-au-lait spots and endocrine dysfunction) (81) and is often operable. The patient may also experience a long survival time with the tumor.

Imaging is useful in the verification of the lesions, particularly for the diagnosis of fibrous dysplasia. Ultrasound examination is safe for the patient and is necessary for the diagnosis of intramuscular myxoma. The typical ultrasound features of MS are considered to be well-defined, hypoechoic masses that are formed from numerous small, fluid-filled spaces that join to form a microcystic pattern (22). CT scans, particularly coronal view scans, are useful in determining the size and quantity of fibrous dysplasia lesions, as well as verifying any bony involvement.

The optimal treatment for MS is surgical excision. Although MS is a benign tumor, it has a tendency to recur, with an accompanying risk of malignant transformation if incompletely excised and commonly requiring 'second-look' surgery (82). The rate of recurrence depends on the surgical approach. The surgical treatment of fibrous dysplasia is mostly indicated where there are progressive deformities, or a risk of fracture or pathological fracture. When the lesions occur in the jaw, the main aim of surgery is tumor resection, with good safety margins, and the reconstruction of the resultant defect, as this is important for the aesthetics and function of the oral and maxillofacial region (83). In the present case, free fibular osteocutaneous flap reconstruction was performed. For myxoma, a total excision is necessary and curative (84). When excluding differentials, such as primary malignancy or metastatic tumors, a full histopathological investigation should be performed for the lesions.

The precise etiology of fibrous dysplasia in association with myxoma remains unclear. The G protein/cAMP/adenylate cyclase signaling pathway that is central to the tissues involved in MAS led to the conclusion that regulatory G $\alpha$  protein (encoded by the GNAS gene) mutations were the underlying molecular etiology of MS (84). This theory is gradually becoming accepted.

In conclusion, clinicians should be aware of Mazabraud's syndrome in order to successfully manage patients with fibrous dysplasia in association with soft tissue myxomas and prevent misdiagnosis of this benign clinical entity as a malignant condition. In addition, differential diagnosis must be continued during clinical treatment when evaluating intramuscular lesions in the context of associated fibrous dysplasia.

## References

- Yaligod V and Ajoy SM: Intramuscular myxoma - a rare tumor. *J Orthop Case Rep* 3: 38-41, 2013.
- Chapurlat RD and Meunier PJ: Fibrous dysplasia of bone. *Best Pract Res Clin Rheumatol* 14: 385-398, 2000.
- Segev Z and Reiner S: Intramuscular myxoma associated with fibrous dysplasia of adjacent bone. *Harefuah* 108: 493-494, 1985 (In Hebrew).
- Arishima Y, Setoguchi T, Abematsu M, Tominaga H, Fukunaga E and Komiya S: Mazabraud's syndrome with solitary myxoma and monostotic fibrous dysplasia. *J Orthop Sci* 15: 144-147, 2010.
- Braunwarth K: Simultaneous occurrence of fibrous dysplasia with extraosseous fibromyxomas. *Fortschr Geb Röntgenstr* 78: 589-594, 1953 (In German).
- Berkhoff WB, ter Brugge Hugenholtz FI and Ingenhous R: A patient with multiple intramuscular myxomas as an extra-osseous manifestation of fibrous dysplasia. *Ned Tijdschr Geneesk* 125: 1460-1462, 1981 (In Dutch).
- Limouzy F, Durroux R, Chiron P, Tricoire JL, Puget J and Utheza G: Fibrous dysplasia associated with intramuscular myxoma. *Rev Chir Orthop Reparatrice Appar Mot* 82: 336-339, 1996 (In French).
- Struk DW, Munk PL and Lee MJ: Musculoskeletal case 8. Mazabraud's syndrome - intramuscular myxoma associated with fibrous dysplasia. *Can J Surg* 43: 15, 62-63, 2000.
- Miyake M, Tateishi U, Maeda T, Arai Y, Seki K, Hasegawa T and Sugimura K: F-18 fluorodeoxyglucose positron emission tomography finding of Mazabraud syndrome. *Clin Nucl Med* 31: 627-629, 2006.
- Zhao H, Zhang H and Jiang Z: Clinicopathological characteristics of Mazabraud syndrome. *Lin Chuang Yu Bing Li Xue Za Zhi* 24: 47-49, 2008 (In Chinese).
- Logel RJ: Recurrent intramuscular myxoma associated with Albright's syndrome. *J Bone Joint Surg Am* 58: 565-568, 1976.
- Henschen F: Fall von ostitis Fibrosa mit multiplen Tumoren in der umgebenden Muskulatur. *Verh Dtsch Ges Pathol* 21: 93-97, 1926 (In German).
- Mazabraud A, Semat P and Roze R: Apropos of the association of fibromyxomas of the soft tissues with fibrous dysplasia of the bones. *Presse Med* 75: 2223-2228, 1967 (In French).
- Zoccali C, Teori G, Prencipe U and Erba F: Mazabraud's syndrome: A new case and review of the literature. *Int Orthop* 33: 605-610, 2009.
- Beele X, Delanote G, Buyse G and Claikens B: Mazabraud syndrome. *JBR-BTR* 91: 84-85, 2008.
- Tang J, Zhao HY, Zheng L, Zhang HZ and Jiang ZM: Abnormal expression of c-myc, p53, p16 protein and GNAS1 gene mutation in fibrous dysplasia. *Zhonghua Bing Li Xue Za Zhi* 38: 292-297, 2009 (In Chinese).
- Yang XD, Xu G, Lü B, Li K and Xu Y: Mazabraud syndrome: Report of a case. *Zhonghua Bing Li Xue Za Zhi* 40: 274-276, 2011 (In Chinese).
- Munksgaard PS, Salkus G, Iyer VV and Fisker RV: Mazabraud's syndrome: Case report and literature review. *Acta Radiol Short Rep* 2: 2047981613492532, 2013.
- Kabukcuoglu F, Kabukcuoglu Y, Yilmaz B, Erdem Y and Evren I: Mazabraud's syndrome: Intramuscular myxoma associated with fibrous dysplasia. *Pathol Oncol Res* 10: 121-123, 2004.
- Kitagawa Y, Ishihara Y, Hayashi M, Kim Y, Fujii N and Ito H: Mazabraud syndrome associated with McCune-Albright syndrome. *J Orthop Sci* 16: 129-132, 2011.
- Lopez-Ben R, Pitt MJ, Jaffe KA and Siegal GP: Osteosarcoma in a patient with McCune-Albright syndrome and Mazabraud's syndrome. *Skeletal Radiol* 28: 522-526, 1999.
- Szendrói M, Rahóty P, Antal I and Kiss J: Fibrous dysplasia associated with intramuscular myxoma (Mazabraud's syndrome): A long-term follow-up of three cases. *J Cancer Res Clin Oncol* 124: 401-406, 1998.
- Krogus A: A case of osteitis fibrosa with multiple fibromyxomatous muscle tumors. *Acta Chir Scan* 64: 465-471, 1928 (In German).
- Uehlinger E: Osteofibrosis deformans juvenilis. (Polyostotische fibroise dysplasie Jaffé-Lichtenstein.). *Virchows Arch Path Anat* 306: 255-299, 1940 (In German).
- Mazabraud A and Girard J: A peculiar case of fibrous dysplasia with osseous and tendinous localizations. *Rev Rhum Mal Osteoartic* 24: 652-659, 1957 (In French).
- Heinemann G and Worth D: Zur Osteofibrosis deformans juvenilis. *Bruns Beitr Klin Chir* 197: 327-336, 1958 (In German).
- Laporte F, Leger H and Malchair G: Albright's syndrome. *J Chir (Paris)* 82: 457-475, 1961 (In French).

28. Lick RF and Viehweger G: A contribution to the diagnosis of fibrous dysplasia of the skeletal system (Jaffe-Lichtenstein-Uehlinger). A roentgenological and pathologic anatomical study. *Fortschr Geb Rontgenstr Nuklearmed* 97: 33-38, 1962 (In German).
29. Roze R, Mazabraud A and Semat P: Fibrous dysplasia of bone and myxomas of the soft tissues. Localized sarcomatous degeneration. *J Radiol Electrol Med Nucl* 48: 527-536, 1967 (In French).
30. Semat P, Roze R and Mazabraud A: Fibrous dysplasia of bone associated with soft tissue myxoma. *J Radiol Electrol Med Nucl* 50: 287-292, 1969 (In French).
31. Wirth WA, Leavitt D and Enzinger FM: Multiple intramuscular myxomas. Another extraskeletal manifestation of fibrous dysplasia. *Cancer* 27: 1167-1173, 1971.
32. Lejeune E, Bouvier M, Vauzelle JL, Queneau P, Thomas JD, Chaudy J, Leung TK and Deplante JP: Fibrous dysplasia and soft tissue myxomas. *Rev Rhum Mal Osteoartic* 39: 281-288, 1972 (In French).
33. Ireland DC, Soule EH and Ivins JC: Myxoma of somatic soft tissues. A report of 58 patients, 3 with multiple tumors and fibrous dysplasia of bone. *Mayo Clin Proc* 48: 401-410, 1973.
34. Sedmak DD, Hart WR, Belhobek GH and Marks KE: Massive intramuscular myxoma associated with fibrous dysplasia of bone. *Cleve Clin Q* 50: 469-472, 1983.
35. Lever EG and Pettigale KW: Albright's syndrome associated with a soft-tissue myxoma and hypophosphataemic osteomalacia. Report of a case and review of the literature. *J Bone Joint Surg Br* 65: 621-626, 1983.
36. Witkin GB, Guilford WB and Siegal GP: Osteogenic sarcoma and soft tissue myxoma in a patient with fibrous dysplasia and hemoglobins JBaltimore and S. *Clin Orthop Relat Res* 204: 245-252, 1986.
37. Blasier RD, Ryan JR and Schaldenbrand MF: Multiple myxomata of soft tissue associated with polyostotic fibrous dysplasia. A case report. *Clin Orthop Relat Res* 206: 211-214, 1986.
38. Biagini R, Ruggieri P, Boriani S and Picci P: The Mazabraud syndrome: Case report and review of the literature. *Ital J Orthop Traumatol* 13: 105-111, 1987.
39. Glass-Royal MC, Nelson MC, Albert F, Lack EE and Bogumill GP: Case report 557: Solitary intramuscular myxoma in a patient with polyostotic fibrous dysplasia. *Skeletal Radiol* 18: 392-398, 1989.
40. Sundaram M, McDonald DJ and Merenda G: Intramuscular myxoma: A rare but important association with fibrous dysplasia of bone. *AJR Am J Roentgenol* 153: 107-108, 1989.
41. Gianoutsos MP, Thompson JF and Marsden FW: Mazabraud's syndrome: Intramuscular myxoma associated with fibrous dysplasia of bone. *Aust N Z J Surg* 60: 825-828, 1990.
42. Prayson MA and Leeson MC: Soft-tissue myxomas and fibrous dysplasia of bone. A case report and review of the literature. *Clin Orthop Relat Res* 291: 222-228, 1993.
43. Gobar GA and Nicholas RW: Case report 800: Skeletal fibrous dysplasia associated with intramuscular myxoma (Mazabraud's syndrome). *Skeletal Radiol* 22: 452-455, 1993.
44. Aoki T, Kouho H, Hisaoka M, Hashimoto H, Nakata H and Sakai A: Intramuscular myxoma with fibrous dysplasia: A report of two cases with a review of the literature. *Pathol Int* 45: 165-171, 1995.
45. Fujii K, Inoue M, Araki Y and Ishida O: Multiple intramuscular myxomas associated with polyostotic fibrous dysplasia. *Eur J Radiol* 22: 152-154, 1996.
46. Court-Payen M, Ingemann Jensen L, Bjerregaard B, Schwarz Lausten G and Skjoldbye B: Intramuscular myxoma and fibrous dysplasia of bone - Mazabraud's syndrome. A case report. *Acta Radiol* 38: 368-371, 1997.
47. Cabral CE, Guedes P, Fonseca T, Rezende JF, Cruz Júnior LC and Smith J: Polyostotic fibrous dysplasia associated with intramuscular myxomas: Mazabraud's syndrome. *Skeletal Radiol* 27: 278-282, 1998.
48. Thomachot B, Daumen-Legre V, Pham T, Acquaviva PC and Lafforgue P: Fibrous dysplasia with intramuscular myxoma (Mazabraud's syndrome). Report of a case and review of the literature. *Rev Rhum Engl Ed* 66: 180-183, 1999.
49. Walker RE, Schwartz RK and Gale DR: Musculoskeletal case of the day. Mazabraud's syndrome (intramuscular myxomas associated with fibrous dysplasia of bone). *AJR Am J Roentgenol* 173: 797, 800-802, 1999.
50. Kransdorf MJ and Murphey MD: Diagnosis please. Case 12: Mazabraud syndrome. *Radiology* 212: 129-132, 1999.
51. Okamoto S, Hisaoka M, Ushijima M, Nakahara S, Toyoshima S and Hashimoto H: Activating Gs(alpha) mutation in intramuscular myxomas with and without fibrous dysplasia of bone. *Virchows Arch* 437: 133-137, 2000.
52. Faivre L, Nivelon-Chevallier A, Kottler ML, Robinet C, Khau Van Kien P, Lorcerie B, Munnich A, Maroteaux P, Cormier-Daire V and LeMerrer M: Mazabraud syndrome in two patients: Clinical overlap with McCune-Albright syndrome. *Am J Med Genet* 99: 132-136, 2001.
53. Delabrousse E, Couvreur M, Bartholomot B, Lucas X and Kastler B: Mazabraud syndrome: A case diagnosed with MRI. *J Radiol* 82: 165-167, 2001 (In French).
54. Pollandt K, Lohmann CH, Werner M and Delling G: Clinical pathological aspects of Mazabraud's syndrome. *Pathologe* 23: 357-360, 2002 (In German).
55. Iwasko N, Steinbach LS, Disler D, Pathria M, Hottya GA, Kattapuram S, Varma DG and Kumar R: Imaging findings in Mazabraud's syndrome: Seven new cases. *Skeletal Radiol* 31: 81-87, 2002.
56. Tsitouridis I, Sayegh FE, Natsis K, Goutsaridou F, Melidis D and Emmanouilidou M: Mazabraud's syndrome: MRI evaluation. *Eur J Radiol Extra* 45: 83-87, 2003.
57. Fang AS, Riley G, Huang SJ and O'Donnell RJA: A 35-year-old man with bony deformities and a soft tissue mass. *Clin Orthop Relat Res* 417: 313-320, 2003.
58. Jhala DN, Eltoum I, Carroll AJ, Lopez-Ben R, Lopez-Terrada D, Rao PH, Pettenati MJ and Siegal GP: Osteosarcoma in a patient with McCune-Albright syndrome and Mazabraud's syndrome: A case report emphasizing the cytological and cytogenetic findings. *Hum Pathol* 34: 1354-1357, 2003.
59. Nguyen BD and Ram PC: Mazabraud's syndrome with sarcomatous transformation: Scintigraphic and radiologic imaging. *Clin Nucl Med* 30: 829-830, 2005.
60. Martin S, Rapariz JM, Osés MJ and Martinez C: A possible cause of multiple intramuscular masses: Mazabraud's syndrome (2007: 11b). *Eur Radiol* 18: 417-421, 2008.
61. McLaughlin A, Stalley P, Magee M, Soper J and Van der Wall H: Correlative imaging in an atypical case of Mazabraud syndrome. *AJR Am J Roentgenol* 189: W353-W356, 2007.
62. Singnurkar A, Phanco JP, Chatha DS and Stern J: The appearance of Mazabraud's syndrome on 18F-FDG PET/CT. *Skeletal Radiol* 36: 1085-1089, 2007.
63. Calisir C, Inan U, Yavas US, Isiksoy S and Kaya T: Mazabraud's syndrome coexisting with a uterine tumor resembling an ovarian sex cord tumor (UTROSCT): A case report. *Korean J Radiol* 8: 438-442, 2007.
64. Macfarlane P, Lew W and Neuhaus S: An aggressive case of Mazabraud's syndrome. *Eur J Surg Oncol* 33: 1087-1089, 2007.
65. Scheepers S, Brys P and Samson I: Case 6532. Mazabraud's Syndrome. <http://www.eurorad.org/eurorad/case.php?id=6532>. Accessed May 8, 2008.
66. Santos CT, Choo CT and Loh AH: Orbital fibrous dysplasia with soft tissue hamartoma - a variant of Mazabraud's syndrome. *Orbit* 27: 207-209, 2008.
67. Tagliafico A, Succio G, Martinoli C and Serafini G: Clinical overlap between Mazabraud and McCune-Albright syndromes. *J Ultrasound Med* 28: 397-399, 2009.
68. Case DB, Chapman CN Jr, Freeman JK and Polga JP: Best cases from the AFIP: Atypical presentation of polyostotic fibrous dysplasia with myxoma (Mazabraud syndrome). *Radiographics* 30: 827-832, 2010.
69. Ijpma FF, Moll FC, Mostert AK, Flach HZ and de Vries JE: Mazabraud syndrome - benign intramuscular myxoma with fibrous skeletal dysplasia. *Ned Tijdschr Geneesk* 155: A2513, 2011 (In Dutch).
70. van der Wal WA, Unal H, de Rooy JW, Flucke U and Veth RP: Fibrous dysplasia of bone associated with soft-tissue myxomas as well as an intra-osseous myxoma in a woman with Mazabraud's syndrome: A case report. *J Med Case Reports* 5: 239, 2011.
71. Gaumétou E, Tomeno B and Anract P: Mazabraud's syndrome. A case with multiple myxomas. *Orthop Traumatol Surg Res* 98: 455-460, 2012.
72. Schimmöller L, Lehwald N, Antoch G and Kröpil P: Polyostotic fibro-osseous lesions associated with intramuscular soft tissue neoplasms. *Radiologe* 52: 934-936, 2012 (In German).
73. John AM, Behera KK, Mathai T, Parmar H and Paul TV: Mazabraud syndrome. *Indian J Endocrinol Metab* 17: 740-742, 2013.
74. Tsourdi E, Hamann C and Hofbauer LC: Skeletal and soft tissue involvement in Mazabraud syndrome. *J Clin Endocrinol Metab* 98: E1381-E1382, 2013.

75. Endo M, Kawai A, Kobayashi E, *et al*: Solitary intramuscular myxoma with monostotic fibrous dysplasia as a rare variant of Mazabraud's syndrome. *Skeletal Radiol* 36: 523-529, 2007.
76. Crawford EA, Brooks JS and Ogilvie CM: Osteosarcoma of the proximal part of the radius in Mazabraud syndrome. A case report. *J Bone Joint Surg Am* 91: 955-960, 2009.
77. Lichtenstein L and Jaffé HL: Fibrous dysplasia of bone. *Arch Pathol (Chic)* 33: 777-816, 1942.
78. Alves C and Silva SF: Partial benefit of anastrozole in the long-term treatment of precocious puberty in McCune-Albright syndrome. *J Pediatr Endocrinol Metab* 25: 323-325, 2012.
79. Adetayo OA, Salcedo SE, Borad V, Richards SS, Workman AD and Ray AO: Fibrous dysplasia: An overview of disease process, indications for surgical management, and a case report. *Eplasty* 15: e6, 2015.
80. Fornage BD and Romsdahl MM: Intramuscular myxoma: Sonographic appearance and sonographically guided needle biopsy. *J Ultrasound Med* 13: 91-94, 1994.
81. He Y, Zhang ZY, Zhu HG, Sader R, He J and Kovacs AF: Free fibula osteocutaneous flap for primary reconstruction of T3-T4 gingival carcinoma. *J Craniofac Surg* 21: 301-305, 2010.
82. Dreizin D, Glen C and Jose J: Mazabraud syndrome. *Am J Orthop (Belle Mead NJ)* 41: 332-335, 2012.
83. Coloma J, García E and Rodríguez A: Intramuscular myxoma. Report of three cases. *Acta Ortop Mex* 28: 244-247, 2014.
84. Walther I, Walther BM, Chen Y and Petersen I: Analysis of GNAS1 mutations in myxoid soft tissue and bone tumors. *Pathol Res Pract* 210: 1-4, 2014.