Primary thoracic chondrosarcoma with intra-abdominal extension in a renal transplant recipient: A case report

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Abstract. Primary thoracic bone tumors are relatively rare. The most common type is chondrosarcoma, accounting for up to 48% of all cases. Patients with primary thoracic bone tumors commonly present with atypical thoracic pain or a solitary palpable chest mass, which gradually develops over months to years. The bones most often affected are the ribs, scapula, costochondral junctions and the sternum. The present study presents a case of a 79 year old previous transplant recipient with a large intra-abdominally expanding chondrosarcoma originating from the left lower thoracic cage and associated vague abdomdinal symptoms. Early recognition and awareness of atypical presentations of this disease are important in to appropriately guide diagnostic evaluation and therapeutic procedures.

Introduction

Primary tumors of the chest wall, arising from soft tissue, bone or cartilage are uncommon. At most, the reported incidence is only 500 new cases per year in the United States (1). Primary bone tumors of the thoracic cage are relatively rare, representing up to 4-8% of all non-secondary, non-metastatic bone neoplasias (2,3). Most chest wall bony tumors demonstrate malignant behavior and the most common primary malignancy is chondrosarcoma (1-4). Waller and Newman (2), using data from the Leeds bone tumor registry, reported that chondrosarcoma constituted around 24% of all bone tumors of chest wall, while other estimates place its incidence at 15-48.1% of the thoracic wall malignancies (4-6).

Chondrosarcomas may arise as *de novo* lesions or in the context of preexisting benign tumors of cartilage origin, such as enchondromas, and are usually located in bones that undergo endochondral ossification (1,7,8). Early recognition, preoperative evaluation and adequate surgical intervention with wide margins are crucial to prevent invasive local disease and metastasis (3,4,8). Awareness of the characteristics and behavior of these tumors are necessary in order to avoid misdiagnosis and inadequate or unnecessary therapeutic interventions (4).

De novo malignancies have become one of the leading causes of late mortality after renal transplantation, with their incidence being 2-15-fold higher than in general population (9). We report a case of large thoracic chondrosarcoma in a kidney transplant recipient with intra-abdominal invasion combined with vague gastrointestinal-related symptoms.

Case report

A 79-year-old woman was referred to our clinic with 3 month history of mild epigastric pain and intermittent nausea, without vomiting episodes. The patient underwent kidney transplantation from deceased donor for end stage renal disease due to diabetic nephropathy 20 years ago, complicated by chronic rejection and return to dialysis the last 5 years. Her immunosuppression regimen included steroids, cyclosporine, and mycophenolate mofetil. Given the clinical presentation and examination findings, a chest and abdomen computerized tomography (CT) scan was performed at an outside facility in order to identify the origin of the palpable mass. Per report, CT revealed a large space-occupying, non-homogeneous solid mass accompanied by intralesional calcifications and measuring approximately 17x15x12 cm (data not shown). In an effort to better characterize the mass and delineate its margins, the patient underwent a magnetic resonance imaging (MRI) scan one week later. It demonstrated a mass arising from the 9th left rib with infiltration of the adjacent subcutaneous fat tissue. Additionally, the mass exhibited intra-abdominal subdiaphragmatic extension with compressive effects on spleen, greater curvature of the stomach, body and tail of pancreas, left colic flexure, left kidney and renal vein. The lesion was well-circumscribed and demonstrated imaging characteristics of neovascularized osteochondral tissue. Preoperative MRI of chest and abdomen did not reveal any suspicious intraparenchymal organ lesion or pathologically

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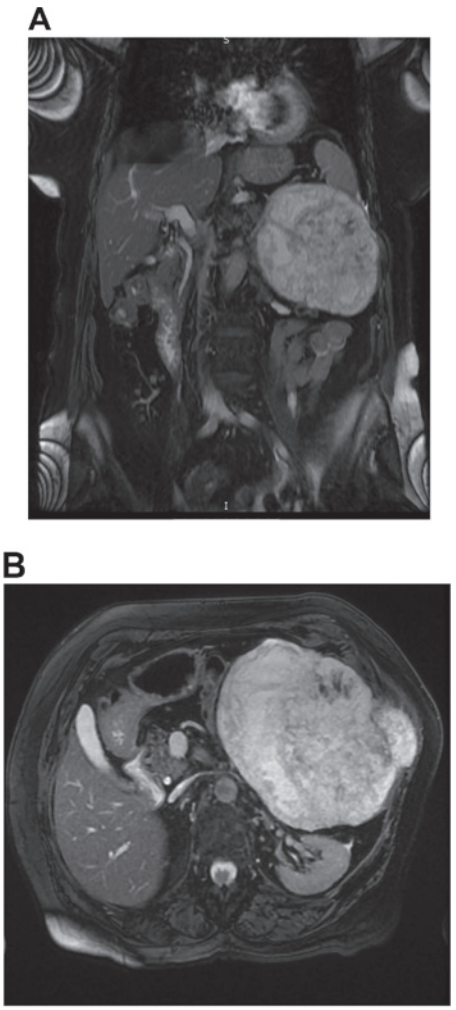

Figure 1. Preoperative coronal and transverse plane MRI. (A) Coronal plane MRI demonstrating the presence of a well-circumscribed, space-occupying, non-homogeneous solid lower thoracic mass with intra-abdominal extension. (B) Transverse plane MRI presenting the intra-abdominal extension of the mass and the absence of intraparenchymal organ lesions.

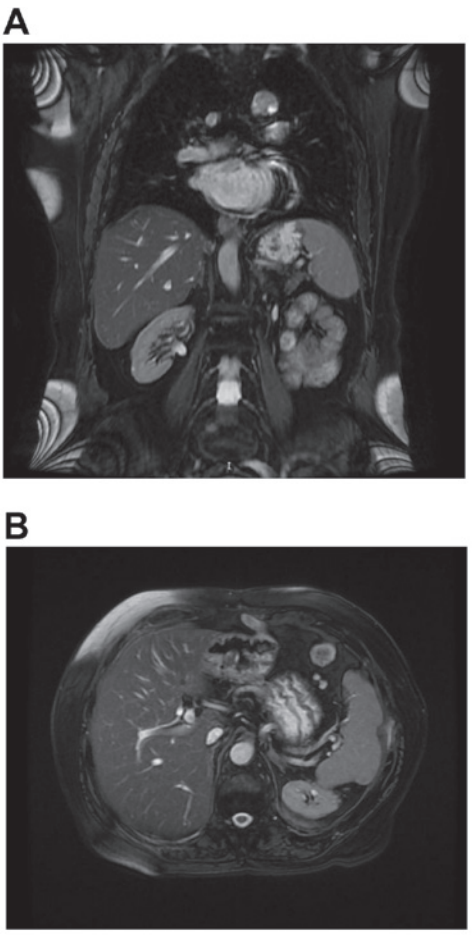

Figure 2. Postoperative 12 month coronal and transverse plane MRI. (A) Coronal plane MRI revealing no signs of local tumor recurrence. (B) Transverse plane MRI revealing absence of tumor recurrence and no signs of intraparenchymal organ lesions.

enlarged intra-abdominal, retroperitoneal or pelvic lymph nodes (Fig. 1).

Considering the clinical presentation and imaging findings, the decision was made to proceed with surgical exploration and resection of the mass with curative intent. Exploratory laparotomy was initiated with a left Kocher incision extended to the xiphoid process and right subcostal area. Intraoperatively, a botryoid, grayish lesion of cartilaginous consistency was identified. Its smooth, membranous external surface was carefully detached from the surrounding tissues with blunt dissection. Despite being in close proximity to the mass capsule, the adjacent organs, such as the spleen, greater curvature of the stomach, pancreas, colon and left renal vein were not invaded. A portion of the diaphragm measuring 12x12 cm was invaded and removed via the abdominal incision. It corresponded to the left costophrenic recess as well as 9 and 10th lateral rib shaft parts.

The diaphragmatic defect was repaired with mesh. The visceral antiadhesive surface was placed adjacent to the left lower lung lobe and the parietal surface put in contact with the spleen in order to avoid adhesion to the small bowel serosa. A left sided chest tube and left subdiaphragmatic drain were also placed. Subsequently, the abdominal wall incision was closed in anatomic layers with simultaneous repair of 2 supraumbilical hernias caused by previous laparotomy. The patient, was transferred to the intensive care unit intubated and hemodynamically stable for monitoring and further postoperative management.

The patient was gradually weaned off the ventilator during the 3rd postoperative day and was subsequently transferred to the surgical ward. Her diet was advanced gradually and the abdominal drain was removed on post-operative day 8, and the chest tube was removed 2 days later. Follow-up radiographic imaging revealed minimal left pleural effusion without any signs of pneumothorax. Histopathologic examination reported the presence of Grade II chondrosarcoma with malignant invasion of the 9 and 10th ribs. The patient was discharged on the post-operative day 14 and referred to medical oncology for further evaluation and management. The patient remained disease-free at the 12 month follow-up, with no evidence of disease on MRI (Fig. 2).

The present study was approved by the Ethics Committee of Laiko General Hospital (Athens, Greece). Data analysis was performed in accordance with the Declaration of Helsinki. Patients who participated in this research had complete clinical data. The signed informed consents were obtained from the patients or the guardians.

Discussion

Soft tissue sarcomas are rare tumors of mesenchymal origin (10). In transplant patients, with the exception of Kaposi sarcoma-which is relatively common with a 400-500-fold increased incidence-soft tissue sarcomas are rarely encountered, thus the diagnostic and treatment algorithms follow those of general population (11-13).

Chondrosarcoma is the most common primary malignancy of the thoracic cage and is usually located in bony tissue developing through the endochondral ossification process (1,3). These malignant neoplasms may arise as lesions in previously healthy bone tissues or develop in preexisting benign chondral tissue tumors, such as enchondromas (1). In addition, chondrosarcomas may develop centrally, in the periphery or on the periosteal surface of the bone where they most commonly affect the ribs, the costochondral junction and less frequently the sternum (1,4,14).

Chondrosarcomas are reported to account for 15-48.1% of the thoracic wall malignancies (2,4-6). Burt *et al* reported that chondrosarcomas of the chest wall correspond to 14.7% of all chondrosarcomas and most of them originated in ribs (43%) and scapula (36%) (15). These tumors predominately affect men with a median age of 50 (1). However, tumors have been reported in patients as young as 11 and as old as 76 (7,16). Risk factors associated with the development of these neoplasms include prior trauma and radiation exposure (often for the treatment of breast cancer or lymphoma), though causation is not well established (1).

Chondrosarcoma grading is an important predictive factor of local recurrence, distant metastasis and patient survival (17). Conventional primary chondrosarcomas are histologically graded as Grade I (low cellularity, abundant matrix, rarely metastasize), Grade II (mitoses present, less chondroid matrix) and Grade III (undifferentiated and highly cellular tumors, with muco-myxoid matrix and mitoses) (17).

Most patients with chest wall chondrosarcoma present with a gradually increasing, palpable, and usually painful mass arising from an anterior chest wall structure such as the costochondral junction or the sternal surface (1). Tumors arising from the anterior chest wall are usually bidirectional (growing outward and inward) and those arising from the posterior thoracic wall tend to grow inwardly only (16,18). Radiographic characterization of these tumors should be the initial step to further guide the appropriate treatment and management (19). Patients presenting with chest wall masses should initially be investigated with a posterior-anterior and lateral thoracic X-rays (1). After this, CT of the chest, including neck area and upper abdomen, is considered as the gold-standard imaging test for accurate tumor localization and preoperative planning (1). On CT, chondrosarcomas characteristically appear as well-circumscribed, lobular masses with intralesional focal, nodular or peripheral calcifications accompanied by adjacent bone or soft tissues invasion (19). MRI does not offer any diagnostic benefit compared to CT and it is not routinely performed in the investigation of suspected thoracic chondrosarcoma (1). In paravertebral, mediastinal or thoracic outlet tumors, where neurovascular invasion is a concern, MRI offers additional information as it offers greater ability to characterizing soft-tissue structures (1). Positron emission tomography (PET) is occasionally used to identify metastatic disease and has also been suggested as a diagnostic modality to differentiate between benign chondral tissue tumors and chondrosarcoma (1,20).

As chondrosarcomas are resistant to radiation and most chemotherapeutic agents, resection considered first line treatment (7,14,16). Surgical intervention involves tumor excision with 3-4 cm margins in order to prevent local disease recurrence (7). However, it is not always feasible to achieve these margins due to proximity to vital neurovascular and visceral structures (7,16). Local tumor recurrence, metastasis, and survival are affected by tumor grade, size, diameter, location and positive margins (7,14,21). Grade I chondrosarcomas have better prognosis compared to Grade II and III tumors, with 10-years survival of 83, 64 and 29% respectively (17). McAfee *et al* reported a higher local recurrence rate in patients treated with excision with negative margins alone compared to a 3-4 cm margin (50 vs. 17%) (21). In addition, 10-year survival dropped from 96% in wide margin group to 65% in local excision (21). The prognosis is more favorable in patients with sternal tumors and tumors measuring <6 cm, whereas survival is <2 years in patients with Grade III tumors (21).

We present a rare case of chest wall chondrosarcoma in a previous kidney transplant recipient, with vague symptoms due to intra-abdominal expansion and invasion. Chest wall chondrosarcomas are rare tumors, especially in transplant populations, which are resistant to chemotherapy and radiotherapy and mainly treated with wide local excision. Patients most commonly present with insidious thoracic pain or solitary chest mass gradually developing over months or years. Awareness of the clinical presentation, appropriate treatment and occasionally atypical progression of these tumors are important to avoid unnecessary diagnostic tests and therapeutic interventions.

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Availability of data and materials

The datasets used and/or analyzed during the present study are available from the corresponding author on reasonable request.

Authors' contributions

DM and SV conceived and designed the present study. DG, DM and SV collected the data. DG and BIS analyzed and interpreted the data. DG, DM and BIS wrote the manuscript. DM, BIS and SV critically revised the manuscript. All authors contributed equally to this work. All authors read and approved the final manuscript.

Ethics approval and consent to participate

The present study was approved by the Ethics Committee of Laiko General Hospital (Athens, Greece). Data analysis was

performed in accordance with the Declaration of Helsinki. Patients who participated in this research had complete clinical data. The signed informed consents were obtained from the patients or the guardians.

Patient consent for publication

Written informed consent was obtained from the patient for the publication of all accompanying images and data.

Competing interests

The authors declare that they have no competing interests.

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