Epidural angiolipoma with concomitant intradural extramedullary capillary hemangioma at the same spinal level: A case report

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Abstract. Spinal epidural angiolipomas (SALs) and spinal intradural extramedullary capillary hemangiomas (SIECHs) are both types of rare benign tumor, and their pathogeneses appear to be associated. The present report is, to the best of our knowledge, the first case of spinal angiolipoma and intradural extramedullary capillary hemangioma occurring at the same spinal level. A 54 year-old male patient experienced two operations within four months due to the occurrence of SAL and one SIECH at the T3 level presenting with sudden paraplegia. Although the co-occurrence of SAL and SIECH at the same spinal level is an extremely rare condition, omitting the intradural tumor may be averted via scrutiny of preoperative images.

Introduction

Spinal angiolipomas (SALs) are scarce benign tumors composed of mature adipose tissue and abnormal vessels, which were first described in 1890 (1). In total, ~200 SAL cases of have been reported in the literature (2). Spinal intradural extramedullary capillary hemangiomas (SIECHs) are more uncommon (3), and to date, only 64 cases of SIECHs have been documented in the literature (4). The oncogenesis of these two tumor types is still unclear, but they may both arise from abnormal primitive pluripotent mesenchymal cells (4,5). Therefore, it is possible that they may appear together. Their clinical presentations are similar, although acute hemorrhage may cause sudden paraplegia (6,7). MRI is the primary method of preoperative diagnosis; however, hematoma may obscure typical features (8). Gross-total resection of all lesions results in a favorable prognosis (9). To the best of our knowledge, the present study is the first case report of SAL with concomitant SIECH at the same spinal level.

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Abbreviations: SALs, spinal angiolipomas; SIECHs, spinal intradural extramedullary capillary hemangiomas; MRI, magnetic resonance imaging; T1WI, T1-weighted image; T2WI, T2-weighted image; CT, computed tomography

Key words: spinal angiolipomas, spinal intradural extramedullary capillary hemangiomas, clinical presentations, diagnosis, treatment

Case report

A 54-year-old male presented to the Outpatient Clinic of Nanfang Hospital due to numbness below the nipples and backache for three days. No injuries were reported. After failure of conservative therapy in a local hospital, the patient was referred to Nanfang Hospital on June 2, 2013. On admission, the patient was already paralyzed with urinary retention. The neurological physical examination revealed T3 level hypoesthesia, 3-4/5 muscle power (Medical Research Council grading) in lower limbs (8), hyporeflexia of knee jerk, negative Babinski's sign and a hypotonic anal sphincter. Emergency MRI was performed, revealing an intradural extramedullary mass located at the T3 level (Fig. 1). The T1-weighted image (T1WI) revealed that the lesion was hyperintense relative to the spinal cord and a T2-weighted image (T2WI) produced an inhomogeneous hyperintense signal. After administration of gadolinium, the lesion was slightly enhanced on T1WI. The initial diagnosis was hemangioma with hemorrhage. Emergency laminectomy and instrumentation were performed revealing an extradural reddish mass with significant feeding vessels. The thecal sac was compressed by the tumor and displaced anteriorly but with clear demarcation. Gross-total tumor resection and coagulation of the feeding vessels were completed. No intradural exploration was performed as the surgeons considered that the size of the resected tumor was consistent with the result of the imaging study and had never previously encountered extradural and intradural tumors in the same location. Postoperative pathological examination revealed mature adipose tissue mixed with a plethora of abnormal vessels of different diameters (Fig. 2). The sample was fixed with 10% formaldehyde overnight at room temperature and embedded in paraffin. The sections (4-μm thick) were stained with hematoxylin for 10 min and eosin for 4 min at room temperature. A light microscope was used to observe the slices. Local hemorrhage was observed and the diagnosis was spinal angiolipoma.

A period of 4 months after the first admission, the patient returned due to uncontrolled backache following treatment

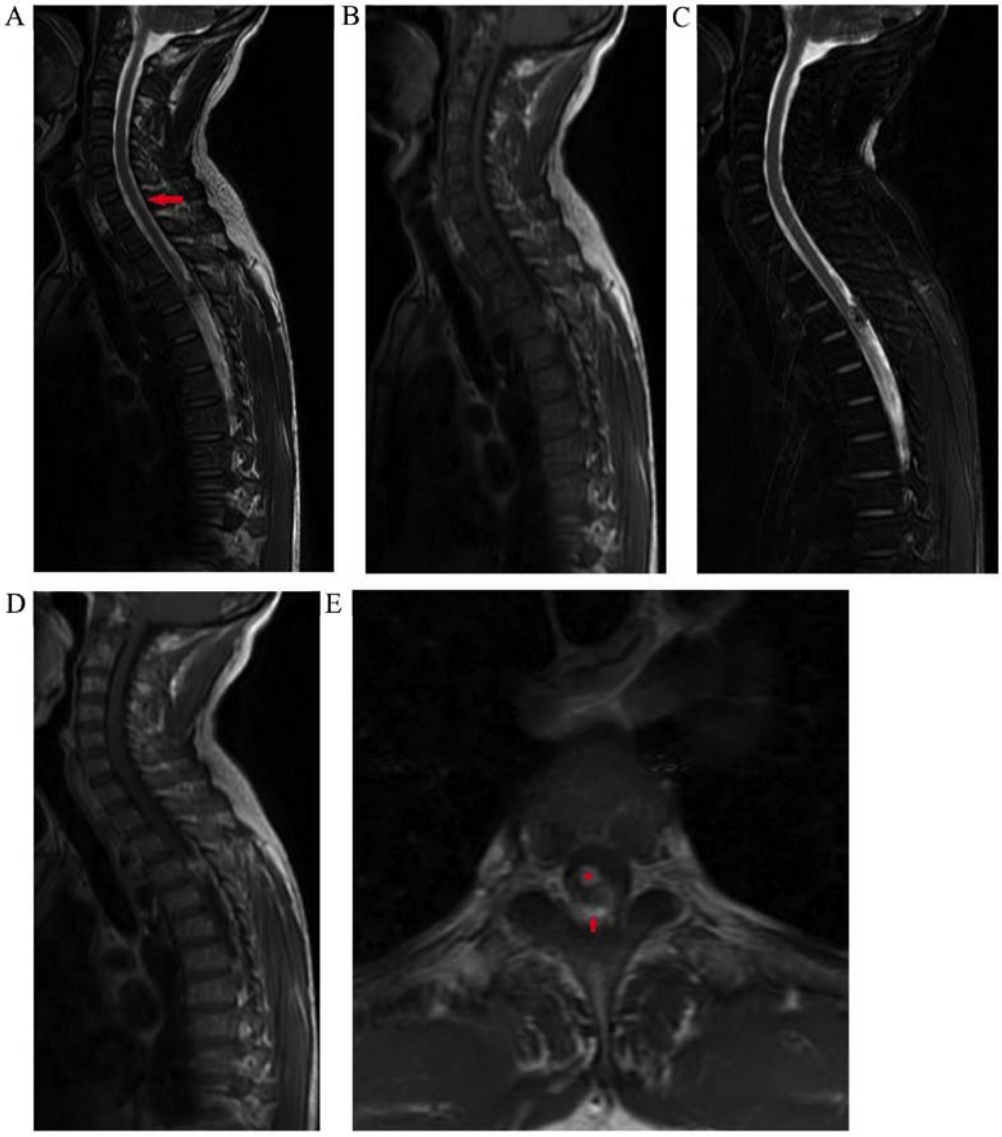

Figure 1. MRI images before the first operation. (A) Sagittal T2-weighted image (TR 3200, TE 124.3). The arrow indicates the linear signal corresponding to dura. (B) T1-weighted image (TR 2860, TE 17.6) and (C) Short T1 Inversion Recovery (TR 3200, TE 124.3) showing an intradural extramedullary mass at T3 level. (D) Sagittal (TR 2860, TE 17.6) and (E) axial (TR 420, TE 12.2; the star indicates the intradural mass, while the arrow indicates the extradural mass) images indicate that the mass is slightly enhanced after administration of gadolinium. TR, repetition time; TE, echo time.

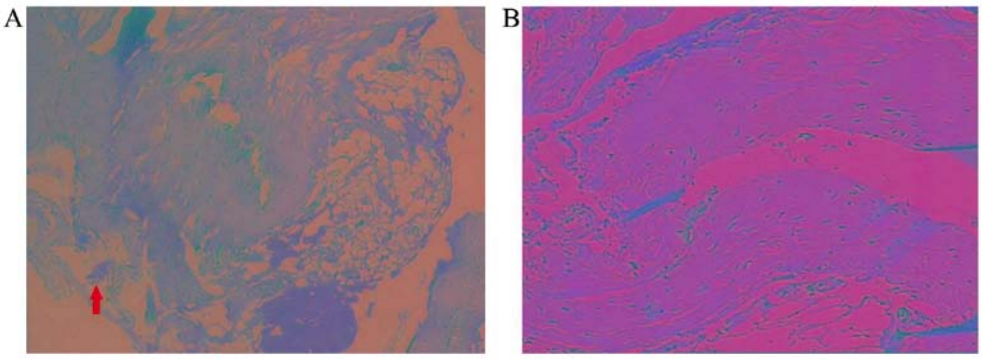

Figure 2. Lesion is composed of mature adipose tissue and abundant abnormal vessels with thick walls, indicating angiolipoma (hematoxylin and eosin staining). (A) Adipose tissue. The arrow indicates tumor hemorrhage (magnification, x20). (B) Abnormal vessels with thick walls (magnification, x80).

with Diclofenac in a local hospital, although numbness of lower extremities had been alleviated and voluntary urination had gradually recovered. All neurological physical examination findings on admission were normal. MRI confirmed a larger intradural extramedullary mass still at the T3 level, with slight hyperintensity both in T1WIs and T2WIs (Fig. 3).

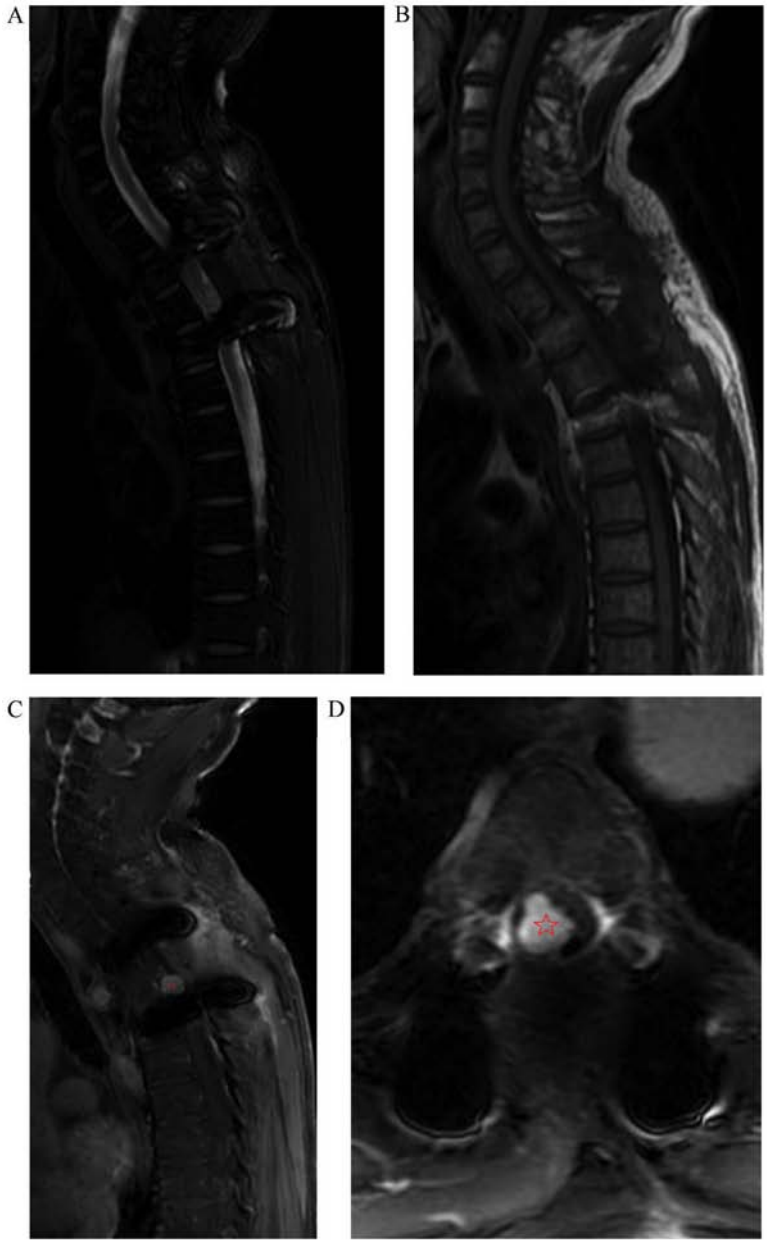

Figure 3. MRI images before the second operation. (A) Sagittal Short T1 Inversion Recovery (TR 2000, TE 98.6) and (B) T1-weighted image (TR 2811, TE 24.1) images display an intradural mass at T3 level. (C) Mass with significant enhancement (TR 150, TE 2.2). (D) Axial T1-weighted image (TR 180, TE 1.8,) indicates that the lesion developed more than 4 months before the first operation. The star indicates the intradural mass. TR, repetition time; TE, echo time.

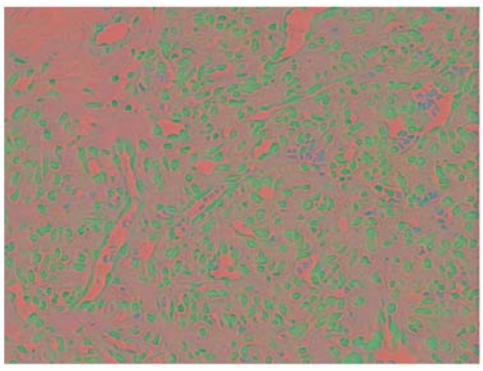

Figure 4. Pathological examination after the second operation reveals hyperplastic fibrous connective tissue mixed with a large number of mature differentiated nascent capillaries. Active proliferation of endothelial cells and infiltration of lymphocytes and neutrophils are also noted (H&E; magnification, x80).

Strong homogeneous enhancement of the intradural lesion was observed after administration of gadolinium, while peripheral soft tissues were slightly enhanced indicating post-operative changes. The second operation was performed via durotomy, revealing a friable, dark-red mass adhering to the pia mater and the enclosure of several neurofibers. The tumor was gross-totally resected. Histologically, the lesion was comprised of mature nascent capillaries with active proliferation of endothelial cells observed. Significant lymphocyte and neutrocyte infiltration were also observed. The diagnosis was juvenile capillary hemangioma (Fig. 4). The patient recovered to normal life 3 months after the second operation. At the 5-year follow-up following the second operation, the patient only presented with mild backache, without any signs of recurrence on enhanced MRI (Fig. 5).

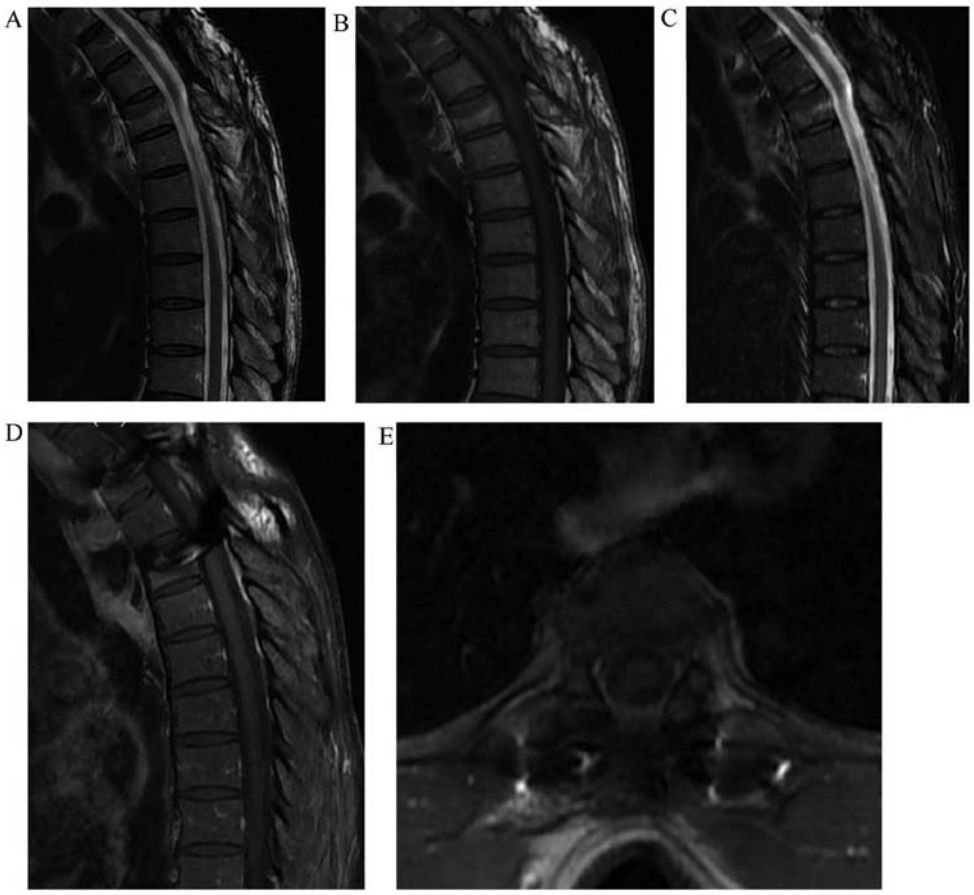

Figure 5. MRI images at 5 years after the second operation. (A) T2-weighted image (TR 2000, TE 222.6), (B) T1-weighted image (TR 385, TE 13.6), (C) Short T1 Inversion Recovery (TR 3402, TE 47.4), (D) enhanced sagittal (TR 376, TE 13.6) and (E) axial T1-weighted image (TR 489, TE 13.2) MRI images indicate no signs of recurrence. TR, repetition time; TE, echo time.

Discussion

SALs are infrequent and only account for 0.04-1.2% of all spinal tumors and 2-3% of epidural spinal tumors (2,9). The incidence of spinal capillary hemangiomas is still unknown, as this tumor is often found in vertebral body, while only <70 cases of intradural extramedullary lesions have been described (4). Both types of tumor are rare and the pathogenesis of these tumors is poorly understood. Certain authors have postulated that SALs may arise from pluripotent mesenchymal stem cells by divergent differentiation along both adipose tissue and angioid lines, similar to a hamartoma (9). SALs may represent an intermediate type between lipoma and hemangioma (10,11), and their vascular components may mimic capillary hemangioma (12). SIECHs were thought to result from impaired movement and differentiation of primitive mesoderm from the embryonic mesodermal plate, also similar to hamartoma (3). Although it is reasonable to assume that they can appear in the same place, to the best of our knowledge, no such case has been previously reported in the literature. In the current article, the first case of the co-occurrence of SAL and SIECH at the same spinal level is presented.

A number of authors have reviewed the demographic characteristics of SALs and SIECHs. SALs predominantly affect patients between 40 and 60 years (1,12), with a slight female predilection (13). The duration of symptoms before presentation may vary from a few minutes to 30 years (1). The most common region affected is thoracic (73.6%), followed by lumbosacral spine (16.9%) (1). On the other hand, SIECHs are more common in males, located in the thoracic-lumbar region and commonly affect patients aged 40-60 years (3,4). Thus, the current case is consistent with the previous literature.

There are different classification systems for both SALs and SIECHs. SALs are categorized into two types: The majority are 'non-infiltrating' and the minority are 'infiltrating', according to whether they involve the vertebra or surrounding tissues (14). The infiltrating type is of either intramedullary or intervertebral occurrence (15). Notably, because 23.8% cases of SALs coexist with vertebral hemangioma (2,12) and the pathology and imaging features of spinal infiltrating angiolipomas with bone involvement are similar to those of aggressive vertebral hemangiomas, it has been questioned whether they represent two distinct entities (6). The classifications of spinal capillary hemangioma type based on 64 patients were as follows: Pediatric (5%), epidural (8%), intradural extramedullary (70%), intramedullary (14%) and hemangiomatosis (3%) (4). In the present case, SAL was classified as the non-infiltrating type due to its clear demarcation from the dura mater and surrounding tissues, while the capillary hemangioma belongs to the intradural extramedullary type. However, given the similar oncogenesis of these two

tumors, it was hypothesized that infiltrating SALs and SIECHs may not represent two distinct entities. Thus, this case may be considered as a subtype of an infiltrating SAL.

The clinical presentations of SALs and SIECHs are similar and do not differ from other benign space-occupying spinal lesions; these include slow and progressive spinal cord compression leading to myelopathy and/or radiculopathy (7). The most common symptom of SALs is paraparesis (30.3%), followed by thoracic/low back pain (24.2%) (1,12). However, rare cases of acute hemorrhage resulting in sudden paraplegia have been documented (16). Only six spinal angiolipomas with tumor bleeding have been reported (16). The current case presents a novel example of the potential adverse effects of SALs. Although the incidence of acute bleeding in SIECHs may be lower than that of cavernous hemangiomas, newly developed hemorrhages within or around the tumor have been reported (7). In the present case report, the acute bleeding which produced sudden paraplegia originated from the SAL rather than the SIECH. Intralesional bleeding of the SAL was confirmed via the conspicuous feeding vessels discovered intraoperatively and the hemorrhage observed in the first pathological study.

MRI is considered to be the most accurate method of investigating SALs and SIECHs. This is primarily because radiographs and computed tomography (CT) can only detect an erosion of pedicles, vertebral body or trabeculation, which are indirect and general signs of intraspinal space-occupying tumors (12,15). Most SALs exhibit hyperintensity on T1WI while the hypointense region on the non-contrast T1WI appears to be of vascular origin, revealing a notable enhancement following gadolinium administration (17). However, the ratio of fat to vessel may differentiate the signal intensity on T1WI (18). Acute hematomas manifest as isointense on T1WI and slightly hypointense on T2WI, while the subacute phase of hematomas appears to be hyperintense on both T1WI and T2WI (17). In the current case, the MRI finding of the SAL pointed to subacute hematoma, although clinically it manifested as an acute process. Unlike SALs, MRI findings of SIECHs are more consistent, showing isointensity in T1WI, hyperintensity in T2WI and homogenous enhancement (19). It was unidentified as to why the SIECH of this case did not exhibit typical MRI features prior to the first operation. It was hypothesized that the blood flow of the SIECH may have been obstructed by the epidural hematoma, as only slight enhancement was observed after administration of gadolinium. After debridement of the hematoma and the SAL, the blood supply of the SIECH was restored to a normal level, which manifested as intense enhancement on MRI before the second operation. Dura mater integrity was confirmed as a hypointense line between the tumor and the cerebrospinal fluid of the subarachnoid space or the spinal cord on T2WI (2), as indicated by the arrow in Fig. 1A. Although the MRI report before the first operation indicated an intradural mass, no further durotomy or investigation for intradural lesions followed, since there was no indication that an epidural and an intradural tumor could exist at the same location. More careful reading of the MRI images, especially the axial enhanced images, would aid in the identification of the two tumors at the same location (Fig. 1E) and help avoid this type of mistake. Human error of the surgeons resulted in the patient receiving two operations within 4 months.

Gross-total resection via laminectomy and durotomy is the primary treatment option for SALs and SIECHs (2,4,6,16). Total removal of infiltrating SALs is impossible in certain cases; however prognosis seems not to differ significantly from completely resected non-infiltrating tumors (20). Angiography and preoperative embolization may be used to diagnose and treat some SALs and SIECHs (20,21). The prognosis of both tumors is good; only a few cases of recurrence have been reported (9,21,22). There were no signs of recurrence of either tumor at a 5-year follow-up in the current case.

In conclusion, spinal angiolipomas and intradural extramedullary capillary hemangiomas are both very rare benign tumors with a similar oncogenesis. Their clinical manifestations are also similar and acute bleeding resulting in sudden paraplegia can occur in both tumors. MRI is the preferred investigative tool for preoperative diagnosis, although typical characteristics may vary due to different cellular composition or formation of the hematoma. Gross-total resection of tumors may result in a favorable prognosis; however attention should be paid to the possible identification of both extradural and intradural tumors at the same level, as in this rare case.

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Availability of data and material

All data generated or analyzed during this study are included in this published article.

Authors' contributions

YC and KL performed the operation, YC wrote the manuscript and HJ analyzed the data and revised the manuscript. All authors have read and approved the final manuscript.

Ethics approval and consent to participate

The present study was approved by the Ethics Committee of Nanfang Hospital, Southern Medical University (approval no. NFEC-201810-K2).

Patient consent for publication

Written consent for publication of the case report and any accompanying images, without any potential identifying information, was provided by the patient.

Competing interests

The authors declare that they have no competing interests.

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