Case Report

Primary mesenchymal chondrosarcoma of the adult lumbar spine: a case report and review of the literature

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Background: Primary mesenchymal chondrosarcoma (PMC) is a relatively rare malignancy that can occur in bone or soft tissue, but rarely in the lumbar spine; there is currently no unified treatment. We report a case of mesenchymal chondrosarcoma originating from the L1 vertebra.

Case Description: A 47-year-old female patient was admitted to the hospital with intermittent low back pain for 20 years, accompanied by intermittent headache and radiating pain in both lower limbs. After admission, magnetic resonance imaging (MRI) showed bone destruction of the L1 vertebral body and accessories and a surrounding soft tissue mass. Enhanced MRI revealed significant enhancement of the L1 vertebral body and soft tissue mass. Technetium 99 m-methylene diphosphonate (99 m Tc-MDP) bone scan showed abnormally high metabolism in the L1 vertebral body, which is highly suspicious of malignancy, and vertebral biopsy revealed a soft tissue malignancy originating from the mesenchymal tissue. Total vertebrectomy combined with postoperative adjuvant radiotherapy was planned, but the patient refused radiotherapy for financial reasons. Intraoperative frozen sections indicated mesenchymal chondrosarcoma, as confirmed by postoperative pathological examination. After 1 year of outpatient follow-up, the patient had no related symptoms, and normal motor and sensory function, and her condition had improved.

Conclusions: Total tumor resection is an effective treatment for PMC, and increased attention to this disease in the clinic is essential.

Keywords: Case report; lumbar spine; mesenchymal chondrosarcoma; biopsy; therapy

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Introduction

Mesenchymal chondrosarcoma is a rare malignant tumor originating from bone or soft tissue (1,2). Morphological histology mainly shows round or ovoid malignant small cells mixed with hyaline cartilage differentiation foci, with a very high malignant potential. Young adults have the highest incidence (2), with a poor prognosis and a tendency for late local recurrence and metastasis (3). There are also reported cases of mesenchymal chondrosarcoma originating from extraskeletal tissues such as the brain and meninges (4). Only five cases of mesenchymal chondrosarcoma originating from the lumbar spine have been reported to date (*Table 1*) (3,5-7). To the best of our knowledge, the patient in this case is the oldest in the published literature. We present the following case in accordance with the CARE reporting checklist (available at https://tcr.amegroups.com/article/view/10.21037/tcr-22-122).

Case presentation

A 47-year-old female with intermittent dull pain in the waist for 20 years, which was aggravated by prolonged sitting or exertion, and accompanied by intermittent

Table 1 Clinical analysis of five cases of PMC of the lumbar spine

No.	Study	Sex/age, years	Tumor location	Histopathology	Immunohistochemistry	Operative treatment	Adjuvant therapy	Outcome
1	Zibis *et al.*, 2010	F/9	L5 vertebral body	Regions of primitive round or spindle-shaped mesenchymal cells alternate with well-differentiated cartilaginous tissue bands	No specific information provided	Staging tumor circumcision surgery; L4-S1 anterior/posterior spinal fusion	Neoadjuvant chemotherapy (vincristine, doxorubicin, cyclophosphamide, ifosfamide and etoposide); postoperative chemotherapy (specific information not provided); image guided radiotherapy: 5,900 cGY in 32 fractions	Alive, no recurrence for 9 years
2	Matsuda *et al.*, 2006	F/44	L1-L2 vertebral body	The tumor tissue differentiated bidirectionally, and small round short spindle-shaped tumor cells and differentiated cartilage islands appear alternately	The tumor widely expressed vimentin and S-100 protein on cartilage island	Total resection of tumor, L1 and L2 vertebral bodies, and tail of T12 vertebral body (including surrounding soft tissues)	Chemotherapy: methotrexate, cisplatin, adriamycin, caffeine	Alive, free of disease after 5 years
3	Fukuda *et al.*, 2019	M/30	L4 vertebral body	Infiltrative malignant biphasic differentiated tumor tissue composed of numerous well differentiated cartilaginous islands and variable numbers of poorly differentiated small cells	The CD99/MIC2 protein was immunopositive in the primary mesenchymal cells. The cartilage region positively expressed S-100 protein, and SOX-9 was expressed in both	L4 total vertebral body resection; vertebral body reconstruction	Systemic chemotherapy (no specific information provided) and local radiotherapy (not specifically mentioned)	Metastasis to scalp occurred 2 months after operation. Skin, muscle, bone and liver metastasis occurred 5 months later
4	Tasdemiroglu *et al.*, 1996	F/12	L5 vertebral body	Mesenchymal chondrosarcoma (no specific information provided)	No specific information provided	L4-L5 laminectomy, gross tumor resection, nerve root decompression, L4-S1 segmental internal fixation	Chemotherapy: cisplatin, epirubicin, ifosfamide; radiotherapy: 5,000 R boost (not specifically mentioned)	Alive, followed up for 10 months, total neurologic improvement
5	Tasdemiroglu *et al.*, 1996	F/1	L1-2 lamina	Poorly differentiated mesenchymal chondrosarcoma	No specific information provided	L1-5 total laminectomy, tumor resection	Chemotherapy: etoposide, ifosfamide, carboplatin, doxorubicin; radiotherapy: 750 cGy (not specifically mentioned)	Alive, followed up for 1 month, the left lower limb was slightly weak and could walk

PMC, primary mesenchymal chondrosarcoma; F, female; M, male.

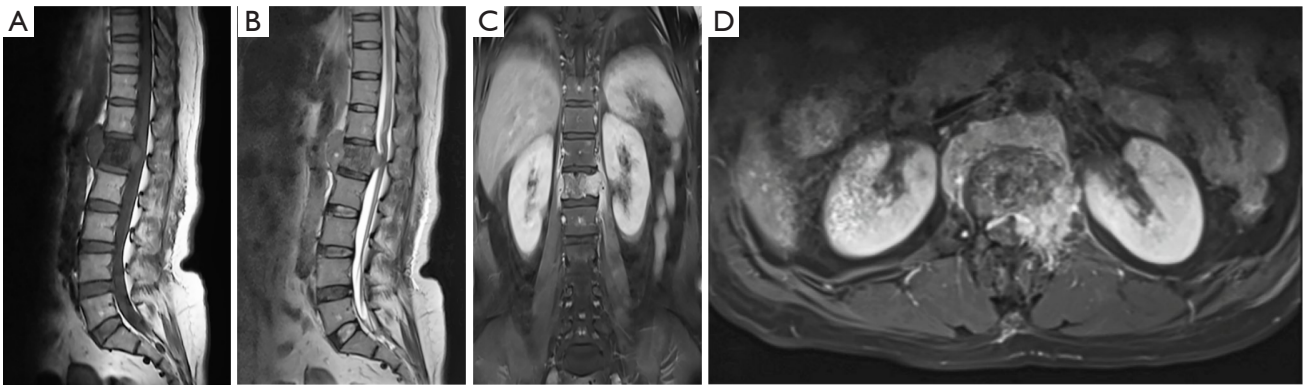

Figure 1 Preoperative MRI of the lesion. Sagittal MRI showed an abnormal signal of L1 vertebra on T1WI (A) and T2WI (B) with the destruction of vertebral bodies and adnexal bone. Coronal (C) and axial (D) contrast-enhanced MRI showed abnormal enhancement signal. MRI, magnetic resonance imaging; T1WI, T1-weighted image; T2WI, T2-weighted image.

headache, limited mobility, and radiation of pain to both lower limbs, was admitted to our hospital for treatment due to exacerbation of the above symptoms in the prior month. Previous treatment was unknown, and there was no related family history or genetic history. Physical examination revealed tenderness to palpation in the lumbar region and restricted movement of the lumbar spine. Quadriceps muscle strength, anterior tibialis muscle strength and gastrocsoleus strength of the lower limbs were all grade 4 bilaterally. The bilateral straight leg elevation test was positive, and the bilateral Achilles tendon reflex was decreased. Laboratory and tumor marker tests indicated no significant abnormalities. Magnetic resonance imaging (MRI) scan showed bone destruction in the L1 and the adnexa, surrounded by a soft tissue mass with low signal on a T1-weighted image (T1WI) (*Figure 1A*) and slightly high signal on a T2-weighted image (T2WI) (*Figure 1B*). Enhanced MRI revealed significant enhancement of the mass (*Figure 1C*,*1D*). Abnormal striped concentration foci in the L1 vertebral body was detected by technetium 99 m-methylene diphosphonate (99 m Tc-MDP) bone scanning (*Figure 2)*. Vertebral bone biopsy under local anesthesia was performed under the guidance of a C-arm X-ray machine to facilitate diagnosis and subsequent treatment. The biopsy showed short spindle-shaped tumor cells with eosinophilic cytoplasm, indistinct cytosolic boundaries and spindle-shaped, ovoid nuclei, which were enriched with branching vessels. Immunohistochemical staining of tumor cells indicated expression of TLE1, INI1, and BCL-2 and partial expression of CD117 and CD99 (*Figure 3*), suggesting a low-grade mesenchymal tissue-derived soft tissue tumor. Because a punch specimen was obtained by biopsy and the phenotype of the tumor could not be clarified, surgical gross total resection was performed.

The patient underwent posterior lumbar surgery with a dorsal median incision centered at the spinous process from T11 vertebra to L3. A layer-by-layer incision of the skin and subcutaneous tissue was made to the lumbar dorsal fascia and through the left and right spinous process, stripping the exposed lamina to the facet joints and transverse process. After determining the entry point for and inserting pedicle screws, L1 total laminectomy was performed. It was found that the L1 left pedicle had been destroyed by the tumor and that the spinal cord and L1 left nerve root were encased in tumor tissue. After careful separation, the T12/L1 and L1/L2 intervertebral discs were completely removed. To prevent local recurrence and metastasis, we use the lower endplate of T12 and the upper endplate of L2 as the boundaries and osteotomized the L1 vertebral body bilaterally; we closely protected the ventral aorta and the anterior longitudinal ligament, carefully separated the surrounding tissues and resected the L1 vertebral body and the surrounding tumor tissue in blocks. Intraoperatively, the spinal cord was seen to be well distended, and no significant compression was detected. An artificial vertebral body was placed in the intervertebral space and adjusted and installed well; the internal fixation was reinforced, and the spinal cord was checked again, with no compression noted. After extensive flushing with saline and tight hemostasis, two drains were placed, the incision was closed layer by layer, and the patient was returned to the ward after stabilization of vital signs.

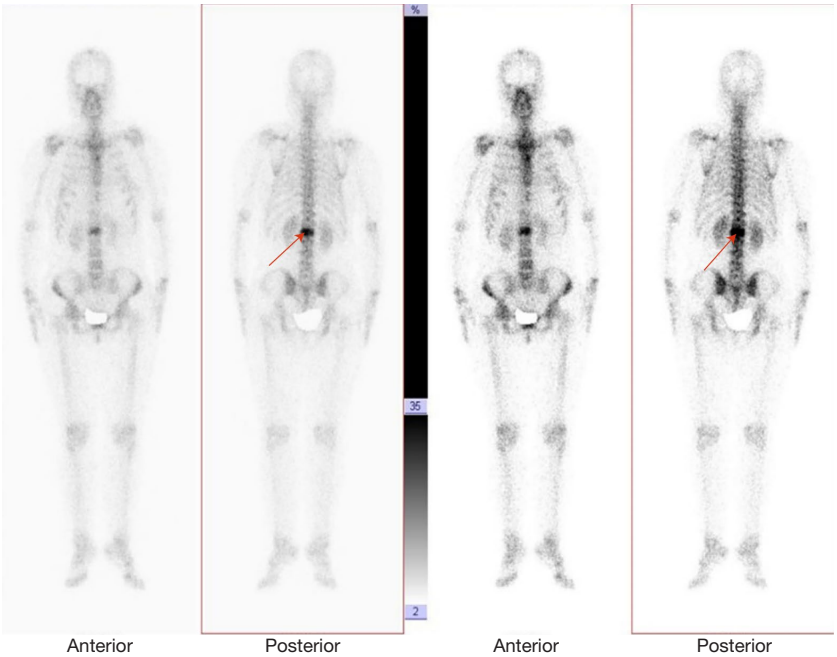

Figure 2 99 m Tc-MDP Whole-body bone scan. Whole-body bone scan reveals abnormal band-like concentration foci in L1 vertebral body with high suspicion of neoplastic lesions (red arrows show). 99 m Tc-MDP, technetium 99 m-methylene diphosphonate.

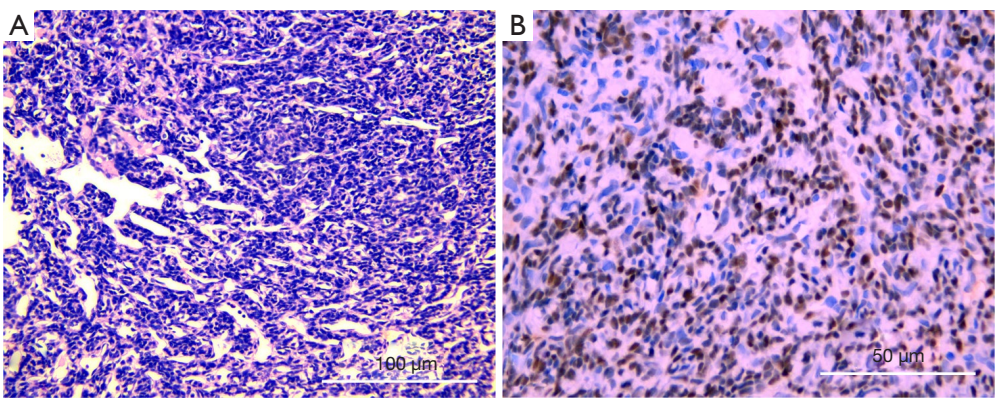

Figure 3 Histopathological examination of vertebral body biopsy specimens. (A) Hematoxylin-eosin showed sections of vertebral body biopsy specimens (original magnification, ×200). (B) Immunohistochemical analysis showed that TLE1 was expressed in vertebral biopsy specimens (original magnification, ×400).

According to pathological examination of the specimen resected, there was no tumor infiltration in the resection margins, demonstrating that the tumor was extensively resected. Postoperative histopathological examination revealed that the tumor tissue consisted of undifferentiated small round cells and insular hyaline cartilage, showing typical bidirectional differentiation. Immunohistochemical staining indicated tumor cell expression of NKX2.2, BCL-2+, SATB-2+, and Ki67 (10%+), partial expression of CD117 and CD99, and no expression of vimentin, S-100, STAT6, H3F3A, MyoD1, Myogenin, SOX-10 (*Figure 4*). These immunohistochemistry results are atypical of mesenchymal chondrosarcoma; in general, mesenchymal chondrosarcomas express S-100 in the cartilage region and vimentin, CD99, and CD57 in small cells (6). Finally, the case was diagnosed as mesenchymal chondrosarcoma

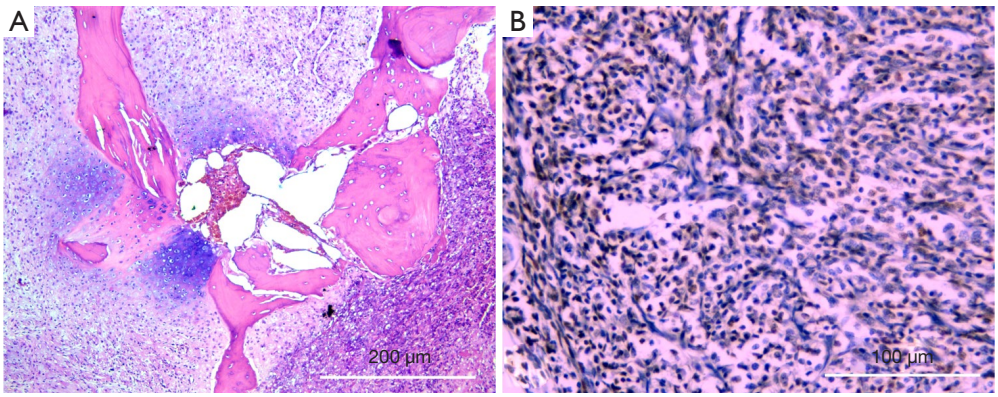

Figure 4 Histopathological examination of postoperative specimens. (A) Hematoxylin-eosin staining of postoperative specimens showed that the tumor tissue showed typical bidirectional differentiation (original magnification, ×100). (B) Immunohistochemical analysis showed that tumor cells express NKX2.2 (original magnification, ×400).

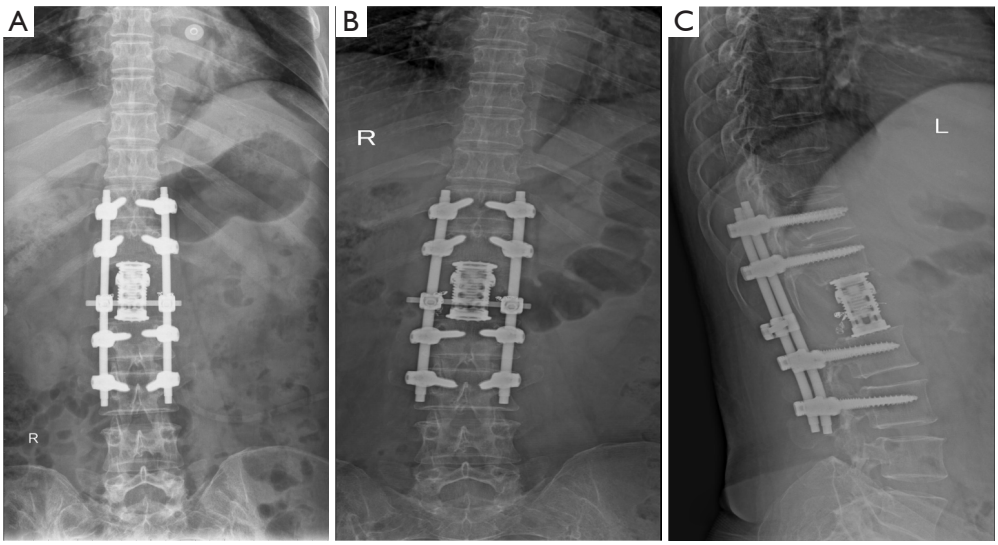

Figure 5 Postoperative and 9-month follow-up X-rays. (A) Postoperative anteroposterior X-rays, and anteroposterior (B) and lateral (C) X-rays after 9-month follow-up. R, right; L, left.

based on various examinations and histopathological findings. After the operation, the patient was stable and could walk on crutches when discharged from the hospital. Radiotherapy was planned after one month, but the patient refused due to financial reasons. During the year of postoperative follow-up, the patient was doing well, was pain-free, showed no corresponding symptoms, and presented no metastasis. Furthermore, the artificial vertebral body and the metal fixator remained in place, with no signs of loosening or slipping on postoperative and follow-up X-rays (*Figure 5*). We will continue to follow up with this patient.

Ethical statement

All procedures performed in this study were in accordance with the ethical standards of the institutional and/or national research committee(s) and with the Helsinki Declaration (as revised in 2013). Written informed consent was obtained from the patient for publication of this case

report and accompanying images. A copy of the written consent is available for review by the editorial office of this journal.

Discussion

Mesenchymal chondrosarcoma is an extremely rare, invasive variant of chondrosarcoma (1,8), accounting for 3% to 10% of chondrosarcomas and 0.2% to 0.7% of malignant bone tumors (9). Lightenstein *et al.* (1) reported the first case of mesenchymal chondrosarcoma in 1959. The majority of cases involve patients in the second or third decade of life, with an average age of 25 years and no sex differences (5). The most common sites of involvement are the maxilla and mandible, but mesenchymal chondrosarcoma may also occur in the spine, ribs, pelvis, humerus, and extraosseous tissues (5). Chondrosarcomas originating in the lumbar spine are rare, as is mesenchymal chondrosarcoma. Hence, mesenchymal chondrosarcoma originating in the lumbar spine is even more rare (1) and worth reporting.

In general, patients with mesenchymal chondrosarcoma seek medical attention due to radicular pain and numbness caused by compression of the lesion. However, a diagnosis of mesenchymal chondrosarcoma is commonly delayed due to the nonspecific clinical manifestations and signs. In this case, the patient had a 20-year history of low back pain. In addition, of the 5 cases found in a literature review, two had a 1-month history of pain at the lesion site (3,7), one had a 9-month history of nausea, lumbar pain, and left flank pain (5), one had a 3-year history of low back pain (6); the remaining case had a 4-month history of back pain (7).

Although imaging examinations may provide a basis for the diagnosis of mesenchymal chondrosarcoma, it is difficult to distinguish it from other types of tumors (9). On X-ray, mesenchymal chondrosarcomas typically appear as a soft tissue mass or osteolytic lesion with indistinct or well-defined borders, and sclerosis is often present. On MRI, most mesenchymal chondrosarcomas are isointense on T1WI and T2WI and tend to be lobulated with well-defined borders (10). The mass in this case was hypointense on T1WI and hyperintense on T2WI, which is quite different from the common isointense manifestations. Overall, imaging examinations are not reliable for diagnosing mesenchymal chondrosarcoma because the imaging manifestations are not specific, and Ewing sarcoma, neurofibroma, and schwannoma should also be considered in diagnosis (9).

Histopathologically, mesenchymal chondrosarcomas are characterized by bidirectional differentiation of tumor cells consisting of alternating small round or spindle-shaped undifferentiated mesenchymal cells and islands of hyaline cartilage (9). The cartilaginous and undifferentiated areas are well defined or intertwined as they migrate, and hemangiopericytoma-like changes can be seen in the undifferentiated areas. Immunohistochemical detection of corresponding biomarkers can help to distinguish mesenchymal chondrosarcomas from other tumors (9). Most mesenchymal chondrosarcomas express vimentin, CD57, CD99, and SOX-9 in small cells and S-100 protein in cartilage regions. SOX-9 is a discriminative marker that is positive in both cartilage areas and small cells and can be used to distinguish mesenchymal chondrosarcoma from other small cell tumors. Nevertheless, it is not a specific marker and can be detected in other cartilaginous tumors (6). The absence of MyoD1 and Myogenin expression can be used to differentiate rhabdomyosarcoma from mesenchymal chondrosarcoma. STAT6 is thought to be a highly sensitive marker for solitary fibrous tumor (SFT), and the absence of STAT6 expression can be utilized to distinguish mesenchymal chondrosarcoma from SFT (11). Overall, diagnosis of mesenchymal chondrosarcoma is difficult, and definitive diagnosis is mainly based on histopathological examination, with immunohistochemistry serving as an auxiliary tool.

As mesenchymal chondrosarcoma has a high recurrence and metastasis rate, treatment should be carefully selected. Kawai *et al.* (12) considered an insufficient surgical margin to be the only independent poor prognostic factor for local recurrence and distant metastasis in patients with chondrosarcoma. In multivariate analysis, surgical margins and tumor grade are statistically significant factors for local recurrence and systemic spread (13). When surgical resection is selected for mesenchymal chondrosarcoma, the extent of tumor resection is reportedly related to overall postoperative survival. At present, complete tumor resection plus stable spinal fixation is the primary choice for treating mesenchymal chondrosarcoma (14). Therefore, total vertebrectomy was performed for our patient to obtain as much total resection of the tumor as possible, and artificial vertebral body replacement was used to enhance spinal stability. Although clinical data and detailed guidelines on the efficacy of radiotherapy for mesenchymal chondrosarcoma are currently lacking, the efficacy of radiotherapy is debatable (9). However, Kawaguchi *et al.* (15) found that postoperative adjuvant radiotherapy reduced the rate of local recurrence. Four of the five patients with

lumbar mesenchymal chondrosarcoma identified in our review received postoperative radiotherapy; one developed scalp metastasis two months after surgery (6), and none of the remaining three developed local recurrence and metastasis during postoperative follow-up (3,7). Thus, we recommend postoperative adjuvant radiotherapy.

Conclusions

Primary mesenchymal chondrosarcoma (PMC) has no specific clinical manifestations and imaging features, and more attention should be given to this disease in clinical work. Furthermore, PMC has a high rate of local recurrence and distant metastasis. Patients should be followed up regularly, and intervention should be actively implemented. Based on the treatment and prognosis of cases to date, total tumor resection combined with postoperative adjuvant radiotherapy is recommended to prevent disease progression and recurrence.

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Footnote

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Ethical Statement: The authors are accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved. All procedures performed in this study were in accordance with the ethical standards of the institutional and/or national research committee(s) and with the Helsinki Declaration (as revised in 2013). Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the editorial office of this journal.

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