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Case Report

Recurrent parachordoma of the lower back: A case report

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ABSTRACT

Parachordoma is a rare entity with less than 50 cases described in the literature. This soft-tissue tumor resembles chordomas as well as extraskeletal myxoid chondrosarcomas and has only recently been fully characterized. Here we describe the case of a patient with a lower back parachordoma and its subsequent postresection recurrence 9 years after the initial procedure, emphasizing the importance of long-term follow-up in individuals with this diagnosis.

Introduction

Parachordomas are well circumscribed tumors with epithelial cells within a myxoid stroma [1]. They were first described in 1951 by Laskowski but until these days only about 50 cases were described in the literature [2,3]. Its true incidence is not known but they seem to be more frequent in young adults and the extremities [1,3]. These are soft-tissue tumors with an indolent nature, slow growth, late recurrences, and rare metastases [1,4,5].

We report a case of parachordoma in the right lower paraspinal area with a late recurrence that was managed with wide resection. To the best of our knowledge this is the second such case to be described in the literature.

Case report

A 36-years-old female presented with a complaint of a mass in the lower right paraspinal area associated with associated intermittent pain. She felt the mass was enlarging in size over the last couple of months. She denied any neurological symptoms radiating down the lower limbs; rather the pain was localized over the mass. As comorbidities, she had hypertension and lupus.

On physical examination a soft-tissue mass of approximately 8 cm by 4 cm was visible and palpable over the right aspect of the lower back. The mass was soft, localized in the subcutaneous tissues, slightly tender, and mobile. No imaging studies were obtained, and the patient was taken to the

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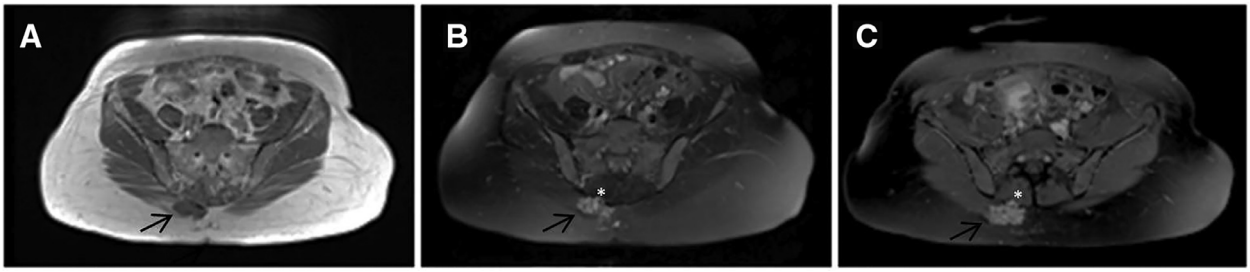

Fig. 1 – Axial MRI images of the lower back at the time of the recurrence. T1-weighted image (A) showing a hypointense lobulated mass (arrow), hyperintense on T2 fat-suppressed sequence (B) and avid enhancement with gadolinium contrast (C). Adjacent muscle invasion is also observed in these images (*).

operating room for a surgical resection. The mass was then sent for pathology assessment and was observed under the microscope as multilobular clusters of neoplastic cells within a myxoid background with some scattered fibrous band; no necrosis or hemorrhage was seen. The sample was sent for immunohistochemical testing as well and stained positively for S100 protein and epithelial membrane antigen. The tumor was reported as compatible with parachordoma and the margins tumor-free.

Nine-years after the initial procedure the patient, now 46-years-old, presented to our center complaining of a new mass in the same area with intermittent moderate pain. Similar to the first occurrence, the patient also felt the mass was growing.

On physical examination, a mass was visible and palpable, below a horizontal scar, along the right lower paraspinal area. The mass was again, subcutaneous, soft, tender, and mobile without pain radiating down the lower limbs nor bladder or bowel abnormalities. A magnetic resonance imaging (MRI) study with and without contrast was obtained. On imaging, a heterogeneous lobulated mass was observed in the right paramidline area. It was iso and hypointense to muscle on T1 sequences, T2 hyperintense, with avid enhancement under contrast and with hypointense septations within. The mass had a focus of extension into the right erector spinae muscle (Fig. 1). It was reported as suspicious for hemangioma or a complex cystic mass. The mass measured $9 \times 4 \times 4$ cm. A core needle biopsy followed with the pathology report indicating findings compatible with parachordoma recurrence.

The patient was taken to the operating room, a longitudinal skin incision was performed over the lesion and electrocautery dissection was carried down towards the lesion, past the fascial plane and down to the sacrum. The tumor was resected en-bloc with a margin of normal-looking tissue around it.

On gross examination, the tumor measured $8.2 \times 4.0 \times 4.0$ cm. It was irregular, white-tanned, with a slight fusiform fibro-fatty surface with pinpoint hemorrhage. Margins were free of tumor. Under microscope examination histological sections revealed nests of epithelioid cells with abundant eosinophilic to vacuolated cytoplasm suspended in a myxochondroid matrix. Mild degrees of nuclear atypia and rare mytotic figures were also observed (Fig. 2). Necrosis was not present. On immunohistochemical testing, it stained positive for CKAE1/3CAM5.2 and S100 protein, it was negative for CK7, 3BetaE12, and Brachyury. Thus, the mass was deemed compatible with parachordoma and matched the prior specimen from 9 years before.

Now the patient is 13 months out of the second procedure with symptoms having completely resolved. The last MRI imaging obtained at 12 months showed no signs of recurrence of the disease. She will continue to follow-up with surveillance imaging studies every 6 months for the first 2 years and then annually.

Discussion

More than 20 years after the initial description, Dabska published a more detailed depiction and a case series of 10 Parachordomas [5]. Only about 50 cases have been described so far with reported cases demonstrating a slight predominance towards male distribution [3]. This tumor occurs more frequently on the fourth decade and in the extremities followed by thorax, trunk, and pelvis. Three back cases have been described, with only 1 reported recurrence occurring 12 years later [5,6,7]. The most common presentation for this rare tumor is nonspecific with most patients presenting with a painless swelling or slow growing mass. Painful masses have also been described, particularly large lesions [8,9].

Other studies have described this tumor on MRI, similar to our case presentation. The tumor was shown to be iso to hypointense on T1-weighted sequences, heterogeneously hyperintense on T2-weighted images and with avid heterogeneous enhancement with contrast [3,10]. It may be very challenging to diagnose this tumor based solely on imaging given it's nonspecific imaging appearance. Parachordomas have also been described in the past on computed tomography imaging as well-defined, homogeneous, or slightly heterogeneous soft-tissue masses without calcification or peripheral fat stranding [11].

Parachordomas are well circumscribed tumors, visualized under the microscope as clusters of epithelioid, glomoid, and spindle cells, in nodules and whorls all surrounded by a myxoid stroma [12]. In this slow growing tumor mitotic figures are rare and there is usually no necrosis or vascular invasion [3].

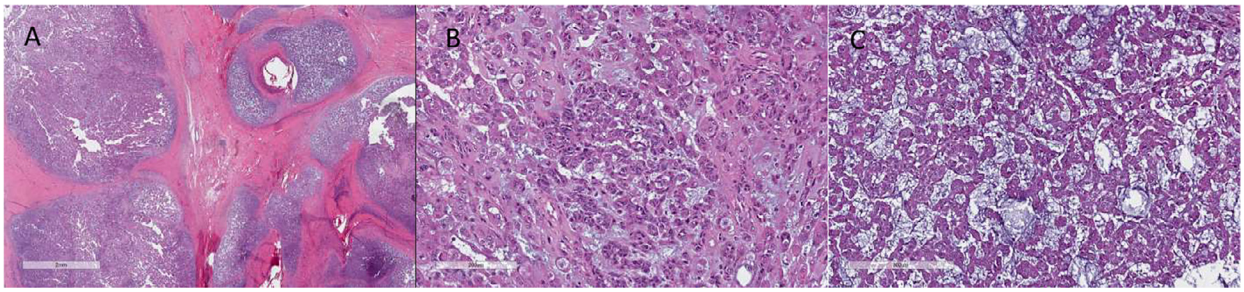

Fig. 2 – * Tumor microscope examination with hematoxylin and eosin stain under different augments (A: 2 mm, B: 200 μm, and C: 300 μm) showing nests of epithelioid cells within abundant eosinophilic and vacuolated cytoplasm suspended in a myxochondroid matrix. *Fig. 2 should be printed in color if available.

Immunohistochemistry is essential to differentiate this tumor from extraskeletal myxoid chondrosarcoma and chordoma. In the former, the cells are usually smaller and they do not stain with cytokeratin (CK) [13]. As for the latter, both tumors stain for S100 protein and epithelial membrane antigen but only chordoma expresses the T-box transcription factor Brachyury [14]. More than 95% of parachordomas stain positive for CK, Vimentin, and S100 protein [1].

Traditionally parachordomas have been considered as benign-behaving tumors, despite the fact that they may recur and even metastasize leading to the death of the patient [1]. Therefore, it is important to achieve negative margins in the resection and continue to follow-up these patients clinically and radiologically for long periods of time, since late recurrences have been known to occur, even 12 years later and in our case 9 years after the initial procedure [5].

Conclusion

Parachordoma is a rare soft-tissue tumor; with an unknown true incidence. Nevertheless, it should be considered in the differential diagnosis of soft-tissue masses, especially in the extremities of young adults. This tumor is recognized for its late recurrences. Thus, long term follow-up may be needed after the surgical resection even when tumor-free margins are achieved given recurrences can present more than 5 years out of surgery.

Supplementary materials

Supplementary material associated with this article can be found, in the online version, at doi:10.1016/j.radcr.2018.09.027.

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