Huge pelvic parachordoma: fine needle aspiration cytology and histological differential diagnosis

Rehab M. Samaka, Mona A. Kandil

Department of Pathology, Faculty of Medicine, Menoufiya University, Shebin Elkom, Egypt

Abstract

Parachordoma is an extremely rare soft tissue tumor of unknown lineage. Parachordoma develops most often on the extremities. Only 2 cases have been reported as pelvic parachordoma. A 46-year old Egyptian woman with a huge painful pelvic mass was found to have a parachordoma with ectopic pelvic right kidney. There is only one report in the literature of fine needle aspiration cytology in this setting. The microscopic picture of parachordoma is not new to pathologists but the gross picture of this rare tumor has not previously been published; not even in the World Health Organization classification of soft tissues tumors. Diagnosis was confirmed by immunohistochemistry. The patient is in good clinical condition without any evidence of recurrence or metastasis after 84 months of follow up.

Introduction

Parachordoma is an extremely rare tumor of unknown lineage that morphologically resembles chordoma of the axial skeleton but occurs in the peripheral site.[1] Since the first report by Lawskowski in 1955, and the later extensive study by Dabaska,[2] only 50-60 cases have been reported in the literature.[3] Parachordoma has a potential for late recurrence up to 12 years.[4] Metastases are even less frequent but a widespread metastasis has recently been reported.[5]

Case Report

A 46-year old woman presented in January 2005 with a slowly increasing abdominal girth and intermittent increasing dull ache pelvic pain of ten months duration. She sought medical advice when symptoms became associated with urinary bladder irritative symptoms. The patient had a past history of mildly enlarged fatty liver, splenomegaly and chronic calcular cholecystitis. All clinical laboratory results were normal. Pelvi-abdominal ultrasonography (USG) and computerized tomography (CT) revealed a large pelvi-abdominal solid mass lesion of hypervascular pattern just deep to the abdominal muscle wall. The lesion abutted the uterus and urinary bladder downward and had a slightly anterior location to the urinary bladder; it was separated from the uterus. The anterior mass location and pattern of displacement suggested a non-adnexial mass lesion. In addition, the right kidney, that remains pelvic, showed normal size and echopattern. No bony lesions could be found. Ultrasound-guided fine needle aspiration (FNAC) and Hematoxylin and Eosin (H & E) stained cytology revealed moderately cellular smears composed of two populations of cell type: epitheloid cells with eosinophilic cytoplasm and spindle-shaped cells in chondroid to myxoid background (Figure 1).

The patient underwent laparotomy. Intraoperatively, the tumor was found to be firmly attached to the posterior aspect of the left rectus sheath and finally to the symphysis pubis. The tumor was compressing but not attached to the urinary bladder, both ovaries and the uterus. Excision of the tumor was performed with the firmly attached left rectus muscle. The excisional biopsy specimen showed a bosselated ovoid, well circumscribed

Correspondence: Rehab M. Samaka, Pathology Department, Faculty of Medicine, Menoufiya University, Shebin Elkom, Egypt.
Tel. +20.1002806239 - Fax: +20.482235680.
E-mail: rehabsamaka@yahoo.com

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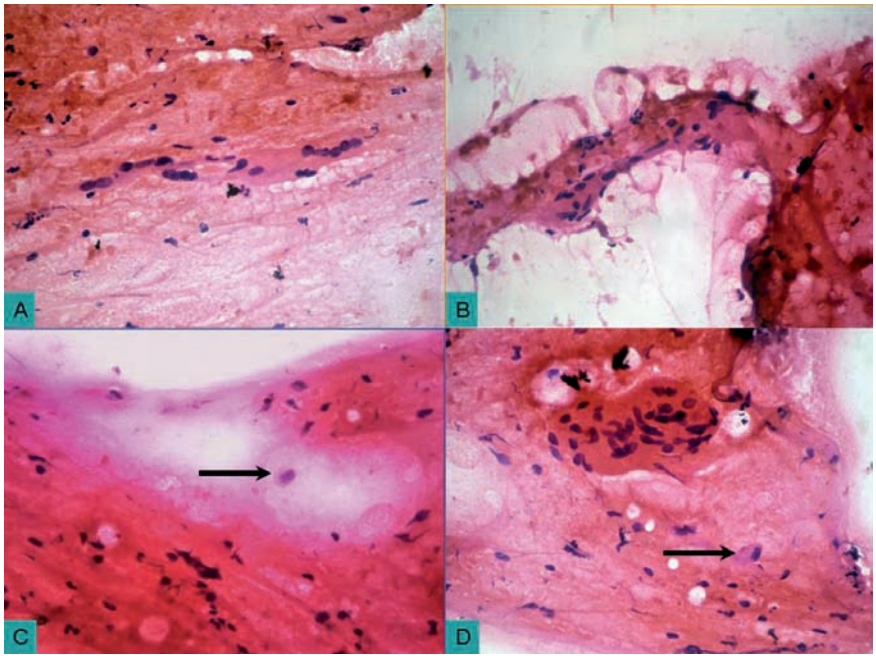

Figure 1. Haematoxylin and Eosin stained ultrasound fine needle aspiration revealed moderately cellular smears composed of two populations of cell type: epitheloid cells with eosinophilic cytoplasm and spindle-shaped cells in chondroid to myxoid background. Plasmacytoid cells are shown (arrows C and D).

pseudo-capsulated soft to firm mass with outer multinodular appearance measuring 15 x 12 x 10 cm. It was attached to the sheathed rectus muscle measuring 11 x 9 cm. The portion of the tumor attached to the rectus sheath measured 5 cm. On the cut surface, there were grayish white fibrous trabeculae irregularly traversing the mass creating the lobulated pattern (Figure 2).

Microscopic morphology displayed well circumscribed pseudo-capsulated neoplastic growth exhibiting irregular bands of fibrous trabeculae traversing the tumor, giving a lobular architecture. The neoplasm was characterized by epitheloid-like cells arranged in small nests, cord-like arrays and a pseudoglandular structure in a myxoid-hyaline stroma. The tumor cells were round and polygonal with abundant eosinophilic to clear cytoplasm which was variably vacuolated, resembling physaliphorous cells. The nuclei were ranged in morphology from a large round structure that had bland vesicular chromatin pattern with a prominent nucleolus to a small pyknotic form. No mitotic figures, necrosis, lymphovascular invasion was observed. Rectus muscle was free from the neoplastic infiltration (Figure 3).

When the periodic acid Schiff reaction was used with diastase - PAS-D, glycogen granules could be found in the cytoplasm of these cells. The tumor cells showed positive immunoreactivity for cytokeratin (CK) AE1/AE3, vimentin, S-100 protein and epithelial membrane antigen (EMA) but were negative for desmin, smooth muscle actin (SMA), HMB-45, Melan-A and cluster of differentiation (CD)-117 (Figure 4). The patient was followed-up for 84 months and was found to be in good clinical condition without any evidence of recurrence or metastasis. The patient and her family gave their consent for the case data to be submitted for publication.

Discussion

This very rare tumor is referred to as extra-axial chordoma, parachordoma or chordoma periphericum.[4] Parachordoma most often develops on the extremities adjacent to tendons, synovium or osseous structures,[5] chest wall,[6] buttocks,[7] skull,[8] hand,[9] and gastric serosa,[10] and has twice been reported in pelvis.[3,11]

Based on the review by Jonathan Clabeaux, there is a slight male predilection in parachordoma, patient age ranges from 4 to 86 years (average age 34.4 years), and the tumor typically occurs in the fourth decade of life in the lower extremity.[3] Our case was female with a huge painful pelvic tumor; this has only been reported twice previously in the medical literature.[3,11] However, the tumor reported here presented with the identical histopathological and immunohistochemical features of the parachordoma, we demonstrated FNAC and the unusual gross morphology. Tumor size was 15×12×10 cm in dimension; 18 cm was the biggest reported in primary intraosseous tumor.[12] Therefore, the tumor in this report is to be considered the largest extraosseous tumor reported in the medical literature.

There is a great diversity of symptoms in different locations but parachordomas are usually painless slowly growing masses.[13,14] However, few parachordomas had localized pain as a presenting symptom, as in our case.[4] To our knowledge, the issue of pain as a symptom in this tumor is poorly defined and has not

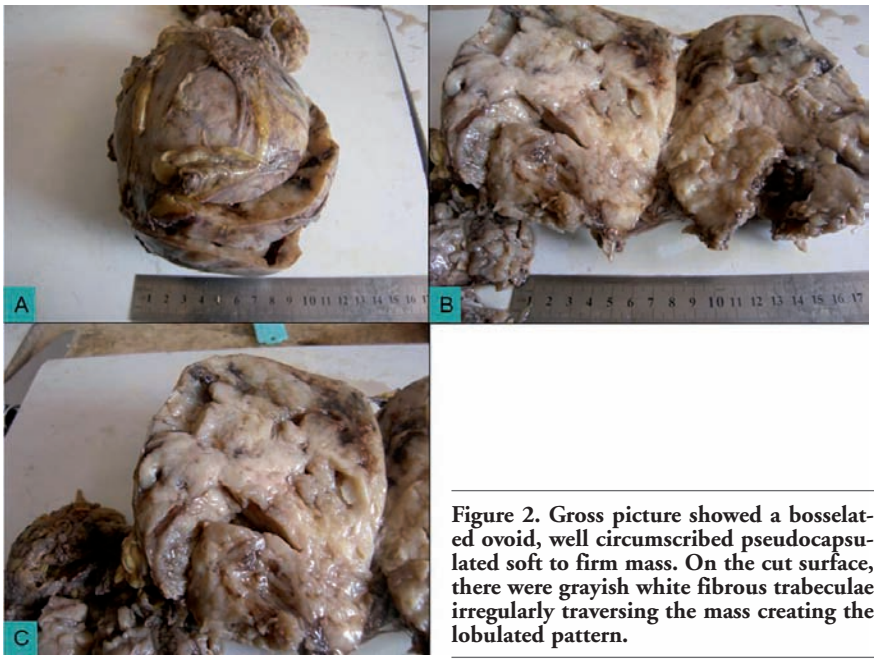

Figure 2. Gross picture showed a bosselated ovoid, well circumscribed pseudocapsulated soft to firm mass. On the cut surface, there were grayish white fibrous trabeculae irregularly traversing the mass creating the lobulated pattern.

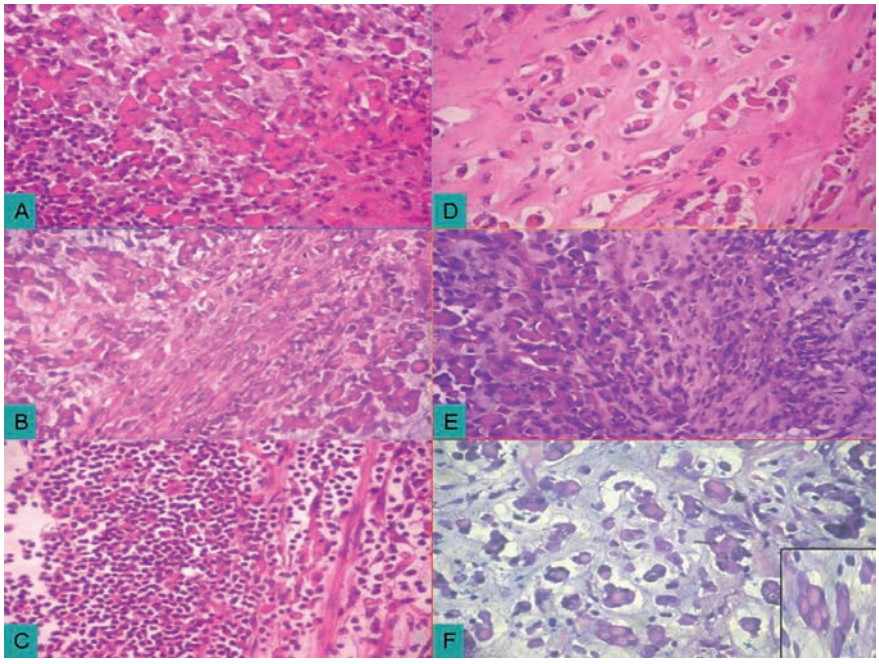

Figure 3. The neoplasm was characterized by epitheloid-like cells arranged in small nests, cord-like arrays and pseudoglandular structure in a myxoid-hyaline stroma (Haematoxylin and Eosin 200×).

been well investigated.

In addition, our case report provides FNAC imaging for this rare tumor, a tumor that has been once recently reported.[15] The histological differential diagnoses for parachordoma from extraskeletal myxoid chondrosarcoma is an important distinction. However, at this unreported site, we must also consider epitheloid/myxoid leiomyosarcoma, extra gastrointestinal tract (GIT) stromal tumor, malignant melanoma and synovial sarcoma as a differential diagnosis because those tumors have a higher risk of local recurrence and distant metastases with histological similarity.

Extraskeletal myxoid chondrosarcoma is distinct from parachordoma in that the cells are smaller and have more intensely eosinophilic but less vacuolated cytoplasm together with a predominant single cell arrangement; they usually do not stain for CK.[16] Epitheloid/myxoid leiomyosarcoma is distinguishable by its immunohistochemical reactivity with SMA.[16] Extra GIT stromal tumor can be excluded by positive immunoreactivity for (CD)-117.[16] Melanomas typically express Melan-A and HMB-45.[16]

In our patient, no local recurrence or metastasis was detected after the 84-month follow up. Parachordoma is considered to be an indolent neoplasm with a potential for late recurrence of up to 12 years.[4] Metastases are even less frequent, but recently a widespread metastasis has been reported.[1] The issue of metastatic potential in this tumor is still poorly defined, and only a few recent reports concerning metastasis of parachordoma have been published so far.[1,17-19] Those metastatic tumors showed both typical and atypical features, such as hypercellularity, pleomorphism and high mitotic rate.[1,18] Consequently, clinical follow up, including a survey of distant metastasis, could be important even if the histological findings show typical features of parachordoma. To our knowledge, there have been only two reports in the medical literature of huge painful pelvic parachordoma. Therefore, this entity could be added to the differential diagnosis list of any painful huge pelvic mass.

To clarify the pathogenesis of pain and its association with the behavior of parachordoma, more clinico-pathological data with closer and longer follow-up information in a larger number of cases is required for this quite rare tumor. More molecular tools must also be used to clarify the picture. These recommendations may help widen our understanding of this enigmatic neoplasm and open the road to new treatment modalities.

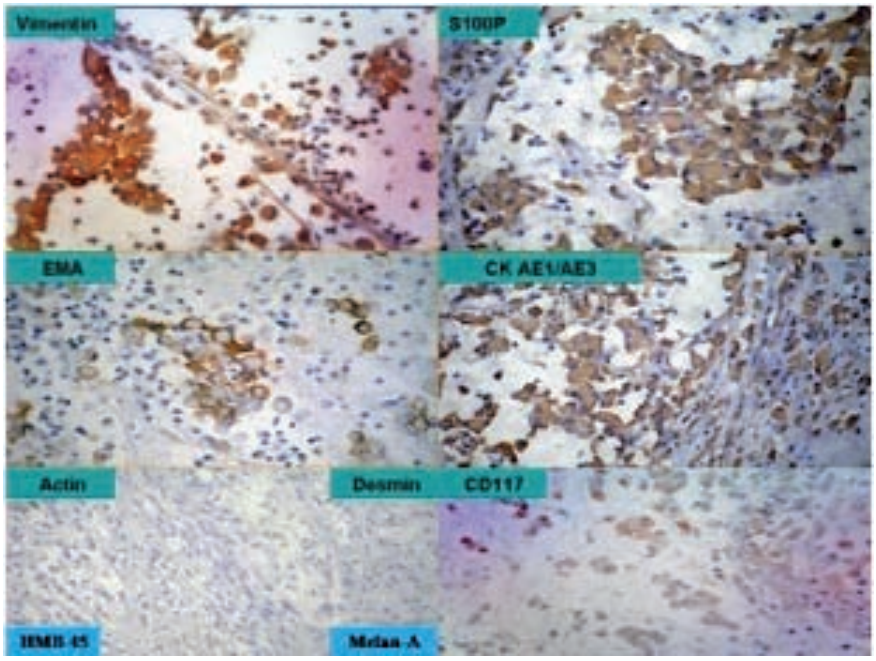

Figure 4. Immunohistochemical study by the labeled antibodies (DAP 400×).

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