Clinical and radiological presentations of pelvic parachordoma

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Abstract

Parachordoma is an extremely rare entity and there are only about 50 to 60 cases reported, in which there is only one definite pelvic parachordoma. We present a huge well-defined presacral tumor in a 48-year-old woman who has the symptoms of lower abdominal pain and difficulty in defecating. Radiological findings of the tumor on computed tomography and magnetic resonance imaging are described in detail. The reasons why we report the case are because the patient has rare clinical symptoms and because this is the second pelvic parachordoma. Then, we summarize the radiological features of parachordoma based on our study and the review of literature.

Introduction

Parachordoma is an extremely rare entity and since the first description by Laskowski in 1951,[1,2] only about 50 to 60 cases have been reported in the literature.[3,4] According to previous studies,[3,5] the majority of the tumors arise from the extremities and the most common symptom is painless swelling. Few cases complain of localized pain. Usually, the lesions on magnetic resonance imaging (MRI) are isointense on T1-weighted images (T1WI) and heterogeneously hyperintense on T2-weighted images(T2WI) with diffuse enhancement.[3,4]

To our knowledge, the case we report is the second pelvic parachordoma,[3] presenting with lower abdominal distending pain and difficulty in defecating. The latter symptom has never been mentioned before. In addition, although the images on MRI of our case are typical, those on computed tomography (CT) are rarely shown before. Finally, combining our article with previous studies, gross radiological appearances of parachordoma are summarized.

Case Report

A 48-year-old woman had complained of mild intermittent lower abdominal distending pain, which was sometimes severe, for one year and difficulty in defecating for two days. Then, she went to our emergency room. Because of her underlying disease of end-stage renal disease, only non-enhanced CT was performed for initial evaluation. On CT, there was an 11cm well-demarcated, slightly heterogeneous (Hounsfield unit: 0-44), and round soft-tissue mass in the presacral region. Fat stranding was not visible and grossly the mass tended to displace rather than to invade adjacent soft-tissue structures, such as the rectum and the uterus. However, since the mass was really huge, some fat planes between the mass and the abovementioned structures are still obscure. Sagittal CT reconstruction revealed that the sacrum was intact (Figure 1). Laboratory data did not show any significant finding except for some abnormal data related to chronic renal failure. Fever was not detected. After admission, this mass was evaluated by sonography, on which it was separated from the uterus and bilateral ovaries. It was heterogeneously isoechogenic with posterior acoustic enhancement and a thin hyperechogenic pseudocapsule. Blood flow was not detected in the mass on color Doppler imaging.

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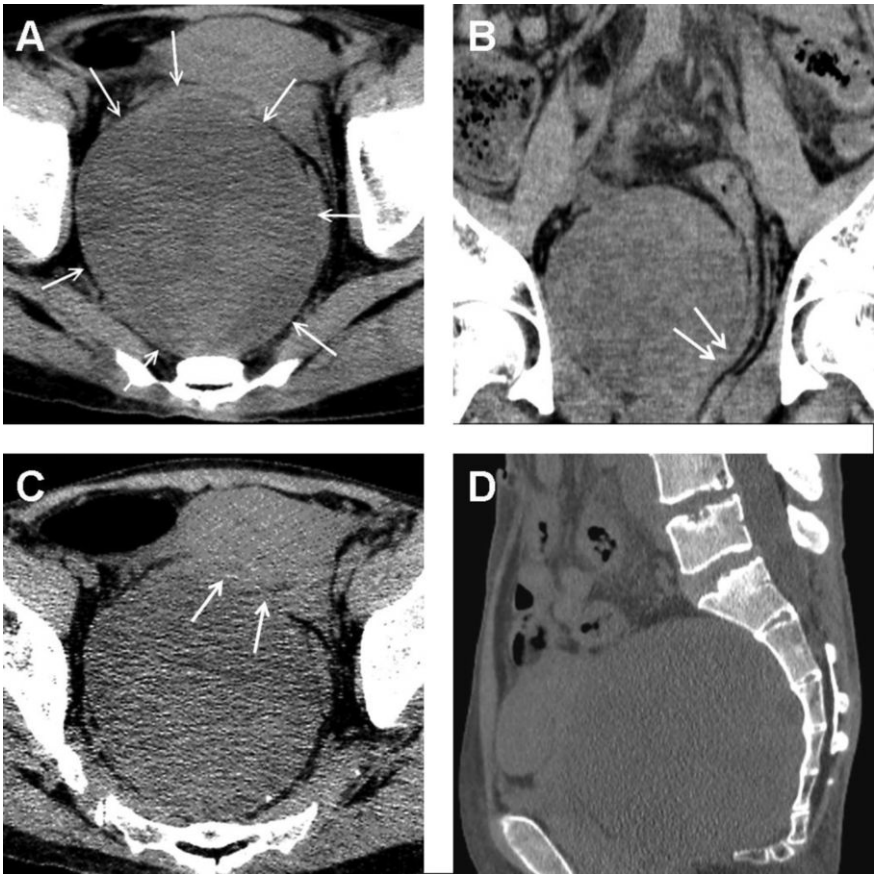

Figure 1. A) Non-enhanced computed tomography demonstrates an 11 cm well-demarcated, slightly heterogeneous, and round soft-tissue presacral mass (arrows) without nearby fat stranding; B) and C) the fat planes between the mass and the rectum (arrows in B) as well as the uterus (arrows in D) are obscure; D) sagittal view in bone window shows that the sacrum is intact.

Owing to the patient's poor renal function, MRI of the lower abdomen was performed only without contrast injection and showed an encapsulated presacral mass, which was isointense on T1WI but heterogeneously hyperintense on T2WI. On fat-suppressed T2WI, there was no signal attenuation in the mass as compared with the images on T2WI. The fat planes on MRI were more clearly identified between the mass and the uterus but still some fat planes were indefinite between the mass and the rectum (Figure 2). Except for this minimally blurred margin near the rectum, this mass was grossly well-demarcated without any soft tissue infiltration toward the skin. After reviewing all the imaging studies in this patient, since the mass was located in the central portion of the pelvic cavity and surrounded by the uterus, bowel loops, and vessels, we chose sonography-guided transrectal needle biopsy as the first invasive procedure for further evaluation. Why we chose the transrectal approach was also because that the mass was almost inseparate from the rectum and that we suspected the mass might be a submucosal tumor of the rectum. Concerning the risk of tumor tract seeding related to a biopsy procedure, we thought the transrectal approach would result in the shortest tract. Afterward, sonography-guided transrectal needle biopsy was conducted and the pathology report favored a mesenchymal tumor with malignant potential. Surgical excision was then carried out and the surgical margin was free from tumor cells. According to the final pathology report, this tumor was composed of epithelioid cells with pale-staining cytoplasm and myxoid or hyalinized stroma. The stroma was stained for alcian blue and was abolished by hyaluronidase predigestion. The arrangement of the tumor cells was in cords, chains or nests (Figure 3). Immunocytochemical stains were positive for Vimentin, S-100, CD99 but negative for epithelial membrane antigen (EMA), Desmin, actin, CK5/6, Calretinin, WT-1, Ber-EP4, alpha-inhibin, and CD34. Type IV collagen embraced groups of tumor cells in a nest-like appearance. Thus, the final diagnosis was parachordoma. This diagnosis was primarily on the basis of cytomorphology and the positive results of Vimentin and S-100. The results of the other immunocytochemical stains were also of some value in this diagnosis. This patient did not receive adjuvant radiotherapy or chemotherapy and there had been no recurrence for three years.

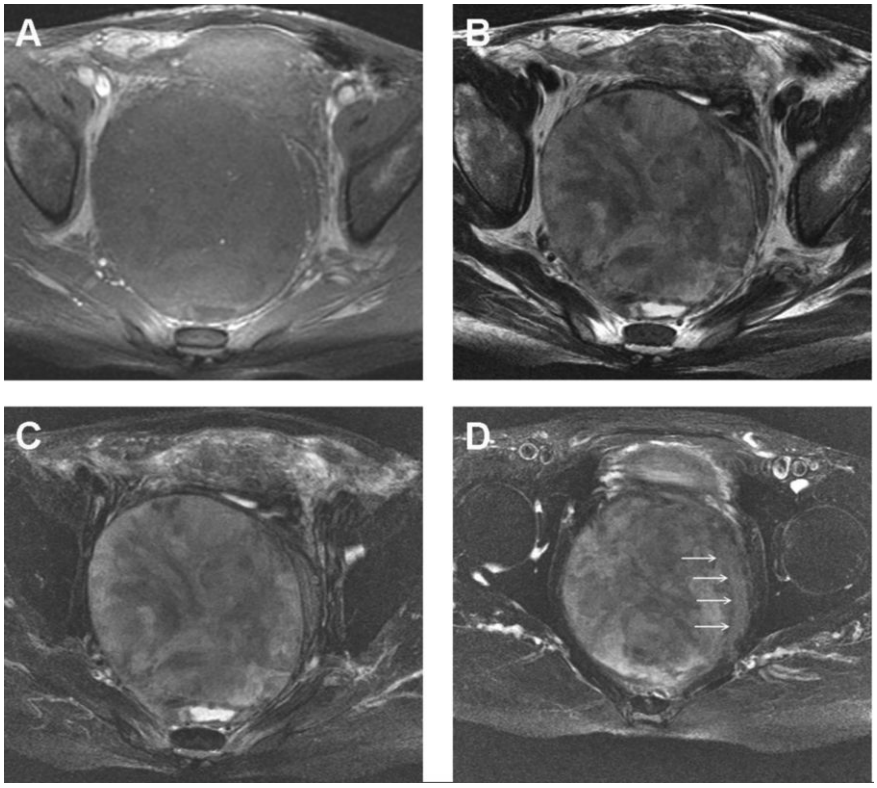

Figure 2. A) T1WI of the lower abdomen demonstrates an encapsulated and isointense to hypointense presacral mass; B) the mass on T2WI is heterogeneously hyperintense; C) signal attenuation is not detectable in the mass on fat-suppressed T2WI; D) still some fat planes are obscure between the mass and the rectum (arrows).

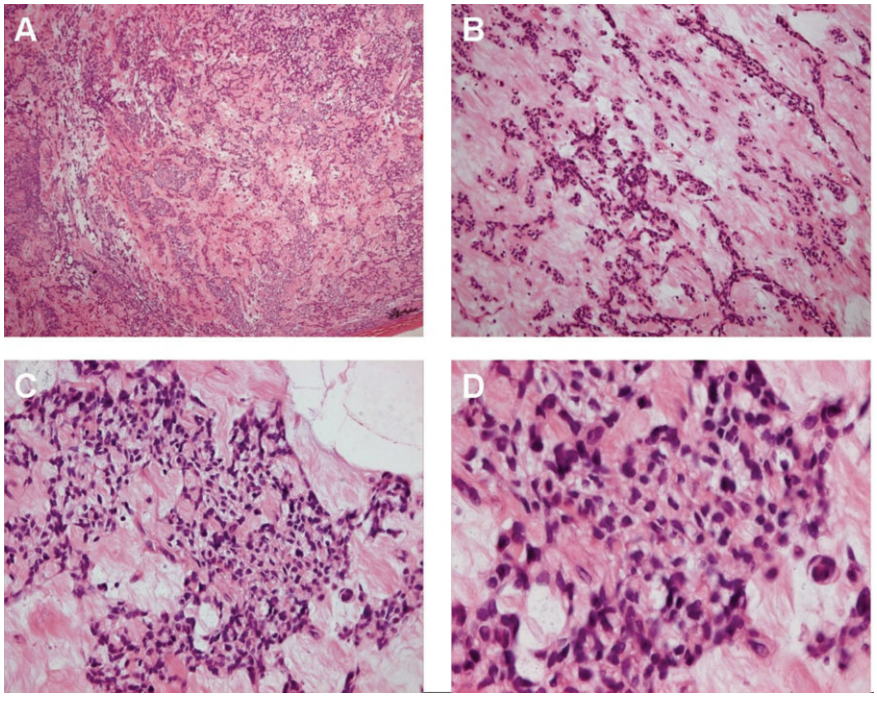

Figure 3. A) The tumor on 20x magnification shows myxoid or hyalinized stroma; B) some tumor cells on 100x magnification form a chain-like arrangement; C) some tumor cells on 200x magnification form a nest-like arrangement; D) the tumor on 400x magnification presents epithelioid cells with vacuolated or pale-staining cytoplasm.

Discussion

Nowadays, parachordoma is considered within the same spectrum with mixed tumor and myoepithelioma.[5] In general, it is a

benign, slow-growing and painless soft-tissue mass; however, recurrence and metastasis have been reported.[3-5] Based on the review by Jonathan Clabeaux, there is a slight male predilection in parachordoma and the age range is from 4 to 86 years with an average age of 34.4 years. The most common location is the extremities (78%), followed by the thorax, trunk, pelvis and nares.[3]

In our review of literature, our case is the second pelvic parachordoma.[3,6] Besides, the symptoms of severe pain and difficulty in defecating are rarely reported. A limitation in our case is that there are no enhanced images on CT or MRI because of her poor renal function. But, combining our case and the review of literature, we can have a more extensive and complete concept about the radiological features of parachordoma, including CT, MRI and even sonography. However, the sonographic appearance of parachordoma is similar to those of many different solid pelvic tumors, and thereby is of low value in diagnosing parachordoma, but sonography may be helpful to survey whether the nearby organs are involved.

On CT, parachordoma is typically a well-defined, homogeneous or slightly heterogeneous soft-tissue tumor without calcification. There is no bony involvement or peripheral fat stranding. The tumor is diffusely enhanced.[7,8] Nonetheless, calcification[9,10] and bony destruction[8,11] occasionally exist. On MRI, parachordoma is hypo- to iso-intense on T1WI and heterogeneously hyperintense on T2WI with diffuse enhancement.[3,8,10] There is no intratumoral fat component or peripheral fat stranding although minimal soft tissue edema surrounding the lesions has been mentioned.[3] The description of the sonographic appearance is mainly based on our study because there is no relevant information depicted in any other study in our review of literature. The mass on sonography is heterogeneously isoechogenic with posterior acoustic enhancement and a thin hyperechogenic pseudocapsule. Blood flow is not detected on color Doppler imaging.

In conclusion, it is obvious that making a correct diagnosis in a case of parachordoma chiefly on the basis of radiological studies is really challenging. However, when encountering a tumor with the abovementioned radiological appearance, we can include parachordoma in the list of differential diagnosis, even though the tumor is not in the extremities, the typical location. The management and prognosis of this benign entity, though some malignant cases exist, are quite different from other diseases with sometimes similar radiological pictures but higher malignant potential, such as chordoma, and some other types of soft-tissue sarcoma.

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