Case Report

Presacral Keratinous and Dermoid Cyst Masquerading as Meningocele – A Rare Case Report

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Abstract

Presacral tumours are rare developmental tumours associated with midline closure defects. A fifteen-month-old girl presented with pain in the lower abdomen and dysuria. After examination, the child underwent investigations and was diagnosed as a presacral mass with intergluteal extension and caudal mass in the subcutaneous tissue of the left gluteal region. Complete excision of mass along with coccygectomy was done. Histopathology showed presacral as a keratinous cyst and gluteal as a dermoid cyst. Later on, the child also developed a left facial dermoid cyst which was also excised.

Keywords: Dermoid cyst, epidermoid cyst, keratinous cyst, presacral tumour

Introduction

In children, various types of masses may affect the presacral area or retrorectal space, i.e. the space located between the rectum and the sacrococcygeal part of the spine. This space contains a variety of tissues such as fat, mesenchymal tissue, lymph nodes, nerve plexuses and vessels. Dermoid tumours are slow-growing cystic masses that are lined by skin with fully mature pilosebaceous structures. Keratinous cysts are common lesions formed by invagination and cystic expansion of the epidermis or the epithelium forming the hair follicle.

Case Report

A 15-month-old girl presented with pain in the lower abdomen and dysuria for 2–3 months. General physical examination was normal. Per abdomen examination did not reveal any abnormality. Local examination showed labial adhesions with a small opening in the vestibule. There was a discrepancy in the size of the gluteal region; the left gluteal region was larger in size as compared to the right with puckering of the skin on its dorsal aspect [Figure 1]. Per rectal examination did not reveal any mass or any abnormality.

The child underwent routine and radiological investigations. Ultrasonography of the abdomen and pelvis showed a cavitary lesion measuring 15.9 mm × 19 mm × 15.1 mm (24 cc) with cystic echogenic collection in the intergluteal region and another lesion on the left side of the buttock measuring 22.2 mm × 19.2 × 20 mm (44 cc). Magnetic imaging resonance was done which showed a well-defined, thin-walled multicystic lesion having fluid-filled signals on all pulse sequences in the presacral space, extending onto the left side and caudally in the subcutaneous region.

Tumour markers alpha-fetoprotein and beta-human chorionic gonadotropin were reported normal. The child was planned for surgery after doing all relevant routine investigations. Complete resection of masses with coccygectomy was done through an abdominosacral approach.

Intraoperative findings showed a 5–6 cm longitudinal, tense, cystic presacral mass which was occupying mainly lower left paravertebral region and was displacing bladder and rectum towards the right side. Another cystic mass of approximately 3 cm × 3 cm was seen in the left gluteal region and was in a proximity to the coccyx [Figure 2]. The clinical diagnosis made was that of presacral teratoma.

Histopathological findings revealed that the presacral tumour had features consistent with a keratinous cyst and the remaining one that under the gluteal region showed the features of a dermoid cyst. No immature elements were present. The

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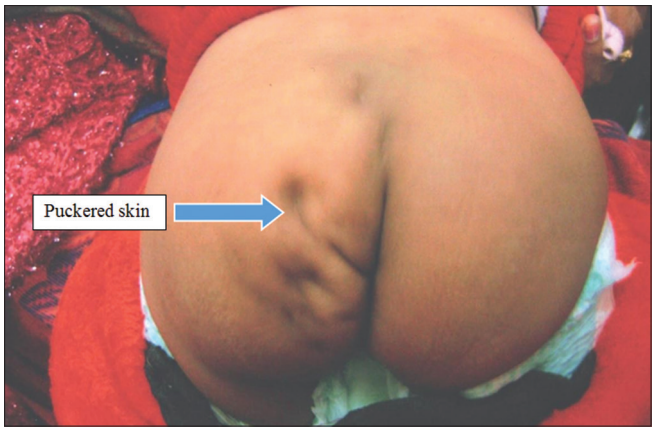


Figure 1: Asymmetry of the gluteal region with puckering on the left side

post-operative period was uneventful. On follow-up, the child developed another small swelling on the left side of the face near the medial aspect of the infraorbital region which was excised and histopathology showed the features of a dermoid cyst.

Discussion

Presacral tumours are rare tumours with an incidence of 1:40,000.[1] The incidence of benign tumours has been reported to be more in females while malignant tumours have a slight male predominance.[2] Caudal embryonic vestiges are postulated to result in dermoid cysts; however, teratomatous lesions are thought to be derived from primordial germ cells which have deviated from a common pathway of development.[3] The presentation of symptoms varies due to the mass effect on adjacent structures or from the pressure effect on the pelvic nerves or bone. The symptoms may include constipation, alteration in bowel symptoms, rectal or sacral pain, pelvic pressure and partial urinary obstruction. The presacral mass may become infected and may present as an infected sinus or abscess.[4] Lack of characteristic symptomatology and difficult anatomical localisation make the diagnostic process difficult and often delay the ultimate diagnosis.[5] Magnetic resonance imaging helps to determine the internal composition of the tumour, whether solid, cystic, fatty or mixed.[6] Dermoid cysts usually occur as solitary lesions; however, multiple concurrent dermoid cysts have also been reported.[7] Epidermoid and dermoid cysts are known to result from defective closure of the ectodermal tube, which results in the inclusion of skin with or without accessory appendages which are lined by stratified squamous epithelium. They are well circumscribed with a thin layer of connective tissue and filled with thick yellow-green fluid containing a mixture of desquamated debris, cholesterol, keratin and water. Epidermoid cysts have no skin appendages whereas dermoid cysts contain them.[8,9] Transabdominal approach whether open or laparoscopically is preferred if lesions are high above the level of S4.[10] In our case, abdominosacral approach was preferred due to its location. A careful dissection away from the rectum should be performed to avoid inadvertent injury to sacral nerves. Due to the paucity of literature about concurrent heterogeneous histopathology as well as anatomical location, there is a need for subsequent research in this field.

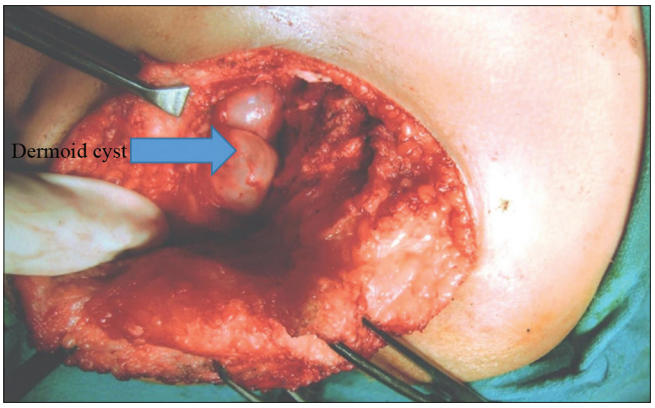


Figure 2: Left gluteal dermoid cyst

Conclusion

The rarity of simultaneous heterogeneous presentation of presacral as a keratinous cyst along with a dermoid cyst on the left gluteal region made us to report this case.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the legal guardian has given consent for images and other clinical information to be reported in the journal. The guardian understands that name and initials will not be published and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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