Case Report

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Congenital Sacral Dermal Sinus – Marker of Presacral Dermoid Cyst

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Abstract

Sacral pits are commonly seen in newborns. They are simple dimples or atypical dimples. Congenital dermal sinus is an atypical dimple and is known to be associated with presacral dermoid cyst. Surgical intervention these without proper evaluation can lead to complications. The management of 8-year-old girl with recurrent congenital dermal sinus is discussed in this case report.

Keywords: Sacral Pits; Congenital Dermal Sinus; Presacral Mass; Dermoid Cyst.

Introduction

Sacral pits or dimples are not uncommon with prevalence of 1-4% quoted in literature [1]. They can be simple or atypical dimples. Congenital Dermal sinus occurs in children in the midline in the sacro-coccygeal region. These dermal sinuses are associated with presacral masses like dermoid cysts and teratomas. Presacral space is a potential space. The masses arising in the presacral space can be divided into following categories congenital, inflammatory, neurogenic, osseous and miscellaneous. Amongst these congenital lesions account for 50% of the masses [2]. Presacral masses are occult lesions with atypical presentations.

Case

A 8-year-old girl had undergone surgical procedures twice at a local hospital for a congenital sinus in the lower back. She presented with complaints of foul smelling discharge from the surgical site. There was no history of constipation. There was a single sinus in the coccygeal region 1 cm above the natal cleft. There was severe scarring of the surrounding tissue (Figure 1). Foul smelling discharge was draining from the sinus. CT fistulogram was suggestive of a pre sacral dermoid cyst with linear opacified tract leading to the sinus (Figure 2). MRI showed a presacral dermoid cyst with hyperintense fat component along with a linear tract extending from the mass up to the skin in the coccygeal region .

Fig. 1: Clinical image showing the scarring around the sinus

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Fig. 2: CT fistulogram

Serum Alfa-feto protein and beta Human chorionic gonadotropin levels were normal. At surgery, the fistulous tract was dissected circumferentially up to the presacral mass. The mass was dissected away from the rectum and excised. Histopathology confirmed the diagnosis of dermoid cyst.

Discussion

Sacral dimples or sacro-coccygeal pits occur in as many as 1-4% healthy newborns [1]. Traditionally these are considered as cutaneous markers of occult spinal dysraphism. Occult spinal dysraphism is associated with sacral dimples in 50-80% of cases [3]. According to Kucera et al only 0.13% of asymptomatic children with sacral dimples required surgical intervention [4]. This brings us to the question which sacral dimple should be evaluated.

Clinical findings alone cannot help us decide with certainty which sacral dimples are associated with occult spinal dysraphism. However, the following findings help us differentiate them into simple dimples and atypical ones. Dimples which are <5mm deep and <2.5 cms from the anal verge and coccygeal pits which are located within the gluteal cleft or oriented caudally are considered as simple dimples. Atypical dimples are >5mm deep and >2.5cms from the anal verge and are associated with one or more of the following, subcutaneous mass, hairy patch, vascular lesions, skin tags or scars, and dermal sinuses (sinuses opening onto skin surface, located above gluteal cleft and have a cephalically oriented tract) [5].

Simple dimples can be safely monitored as long as they as asymptomatic [5]. All symptomatic and atypical dimples should be evaluated with Ultrasonography within the first three months of life, in older infants and children MRI is considered as the best diagnostic tool [6].

Congenital dermal sinus is a tract lined by stratified squamous epithelium occurring anywhere from the nasion to the coccyx. Excluding dimples in the sacrococcygeal region, the incidence of CDS appears to be approximately 1 in 2500 to 3000 live births. Dermal sinuses in the lumbosacral region or higher occur during the process of neuralation whereas those in the coccygeal region occur during the process of canalization of the tail bud. Since a dermal sinus contains dermal components one can anticipate it to be associated with a dermoid cyst [7].

Table 1: Classification by Uhlig & Johnson

Congenital-	Neurogenic	Osseous	Miscellaneous	Other
Benign Developmental cysts (teratoma, epidermoid, dermoid, mucus-secreting) Duplication of rectum Anterior sacral meningocele Adrenal rest tumor *Malignant* Chordoma Teratocarcinoma	*Benign* Neurofibroma Neurilemoma (schwannoma) Ganglioneuroma *Malignant* Neuroblastoma Ganglioneuroblastoma Ependymoma Malignant peripheral nerve sheath tumors (malignant schwannoma, neurofibrosarcoma, neurogenic sarcoma)	*Benign* Giant-cell tumor Osteoblastoma Aneurysmal bone cyst *Malignant* Osteogenic sarcoma Ewing's sarcoma Myeloma Chondrosarcoma	*Benign* Lipoma Fibroma Leiomyoma Hemangioma Endothelioma Desmoid (locally aggressive) *Malignant* Liposarcoma Fibrosarcoma/malignant fibrous hisiocytoma Leiomyosarcoma Hemangiopericytoma Metastatic carcinoma	Ectopic kidney Hematoma Abscess

The presacral space is located between rectum and the sacrum. This place is embryologically significant as it is a place of embroyological crossover of the neuroectoderm, hindgut, and notochord. This gives rise to various tumors and masses in this area. Presacral tumors are rare and the reported incidences range between 1 in 40,000 to 63,000 hospital admissions [8,9].

Uhlig and Johnson first classified these masses in 1975. The modified classification was put forth by Dozois et al who further subdivided the masses into benign and malignanat (Table 1). Congenital lesions are the commonest amongst these lesions accounting for 55 to 70% of all lesions [10].

Dermoid and epidermoid cysts result due to abnormal ectodermal tube closure. Epidermoid cysts are lesions composed of stratified squamous cells. These are usually benign and unilocular. Dermoid cysts in addition to stratified squamous epithelium contain skin appendages like sweat glands, hair follicles, or sebaceous cysts and thus can be differentiated from epidermoid cysts. Both these type of lesions may be associated with a postnatal dimple or sinus when they communicate with skin [10].

These masses due to their location present with non-specific symptoms. Thus, in presence of a dermal sinus a thorough evaluation of abdomen and pelvis by Ultrasonography is warranted. Surgery is mainstay therapeutic option. Complete excision of the mass along with the sinus tract ensures complete cure.

Conclusion

Sacral dimples are not uncommon in healthy term newborns. Atypical dimples should be evaluated with USG and MRI, as they are markers of sinister internal pathologies. Congenital dermal sinuses are associated with presacral dermoid cysts and hence should be thoroughly evaluated before surgery.

Conflict of Interest: Nil

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