Median Raphe Cyst: A clinically challenging diagnosis.

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Abstract

Median raphe cyst is an uncommon developmental anomaly that can develop anywhere along the midline of the external genitals. Only a few hundred cases have been published in the English literature and the lack of awareness of this entity can lead to confusion and misdiagnosis. We report here a case of median raphe cyst located in the median of the anterior scrotum of a man in his mid-thirties. The clinical presentation was that of a scrotal mass increasing substantially in size over two days associated with tenderness, skin erythema, and scrotal pain. Radiologic interpretation of a sonogram and computed tomography scan suggested a thrombosed vessel. The patient was diagnosed with septic thrombophlebitis associated with overlying cellulitis. Despite conservative therapy with antibiotics, the patient developed pyrexia, tachycardia, and leukocytosis prompting surgical excision of the lesion. Histopathologic examination revealed an infected median raphe cyst. The cyst wall was lined by a stratified epithelium that included numerous Alcian blue positive goblet cells. The epithelial cells showed reactive changes with infiltration by numerous neutrophils. Our objective is to bring attention to and thereby facilitate the diagnosis of this unusual entity.

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Introduction

Median raphe cyst is an uncommon anomaly that can develop at any location over the midline of the external genital region from the anus to the scrotum and perineum. This uncommon cystic lesion was first described by Mermet in 1985.[1] Since then, a few hundred case reports have been published in the English literature describing cysts located over the glans penis, penile shaft, perineum, para meatus and scrotum. We are reporting a new case of median raphe cyst located over the anterior scrotum of a male in his mid-thirties presenting as a painful scrotal mass.

Case report

A male patient in his mid-thirties presented to the Emergency Department (ED) with scrotal pain. History goes back to approximately two years prior to this presentation when he first noticed a painless scrotal mass. During that time, the mass has slowly grown. It has never caused him pain or other symptoms. In the 2 days prior to seeking medical advice, the lesion became increasingly larger and more painful with a sense of fullness, as described by the patient. In addition, the overlying skin became red and painful to touch. The patient underwent a sonogram and CT scan that suggested a thrombosed vessel. No history of urologic trauma or infections were reported. The patient was diagnosed in the ED with septic thrombophlebitis associated with overlying cellulitis. He was started on Vancomycin, Clindamycin and Rocephin. The patient failed conservative therapy with antibiotics and started developing a fever of 102.6F, tachycardia and leukocytosis. General surgery was consulted and they decided to remove the lesion surgically in the operating room. The scrotal lesion was removed and sent to pathology to reveal an infected median raphe cyst.

Imaging Studies

Sonography:

Fig.1. Ultrasound showing the 7.5 x 1.5 x 1.2 cm avascular cystic structure.

Left hemiscrotum: Testicular morphology and flow is normal, 4.6 x 2.7 x 3.2 cm. There is a small hydrocele, the epididymis is normal. Along the posterior scrotum, extending from the base of the penis at least 8 cm is a 7.5 x 1.5 x 1.2 cm avascular fluid filled tubular structure with thickened walls. Radiologic Impression: 1. Small bilateral hydroceles, normal testicular morphology, 2. Unusual complex tubular structure superficial posterior scrotum, no associated mass, most likely occlusion of superficial vein.
CT scan Abdomen and Pelvis:

Fig. 2. CT scan showing the scrotal cyst along the scrotal midline.

Reproductive organs: Concordant with ultrasound is an avascular appearing tubular structure that extends from the base of the penis anterior and inferior along the scrotum. No overt findings of infection in the regional fat. Radiologic Impression: Avascular tubular structure superficial scrotum as noted above, most likely thrombosed vessel, consider urologic consultation.

Microbiology

Culture of the fluid found in the cystic lesion revealed rare Staphylococcus species, coagulase negative in 2 colony types.

Pathology Report

Final diagnosis:

Median raphe cyst with abscess.

Microscopy:

Sections demonstrate a cyst wall lined by a stratified epithelium that includes numerous Alcian blue (Fig. 3) and Mucicarmine (Fig. 4) positive goblet cells. The Alcian blue/PAS and Mucicarmine stains included concurrently performed controls showing appropriate reactions.
The epithelial cells show marked reactive changes with infiltration by numerous neutrophils. The surrounding connective tissue shows hemorrhage, edema, and acute and chronic inflammation with a prominent neutrophilic infiltrate. Squamous differentiation is present in some foci (Fig. 5 and Fig. 6).

Other sections showed an abscess just deep to the epidermis focally lined by flattened epithelium similar to that described previously, but which had been denuded in many areas and attenuated in others (not shown).

Figure 3. Alcian Blue / PAS stain x400
Figure 4. Mucicarmine stain x400
Figure 6. Transitional epithelium mixed with goblet cells. H&E x200
Figure 5. Squamous epithelium lining. H&E x200
Discussion

Median Raphe cysts are a benign cystic lesion generally present at birth [2]. They can develop at any location over the midline of the external genital region from the anus to the scrotum and including perineum [1]. They are commonly diagnosed in patients less than 30 years of age [3]. These lesions were described and reported under different nomenclature in the past including urethroid cyst, mucoid cyst, genitoperineal cyst, apocrine cystadenoma and hydro-cystadenoma [2]. Although most are asymptomatic or unnoticed during childhood, they are more likely to produce symptoms if they are located distally [4]. They may grow slowly and become symptomatic, causing pain, swelling and erythema due to secondary infections or trauma [5]. In our case the patient presented with a swollen, painful and erythematous scrotal lesion diagnosed as thrombophlebitis by imaging studies. Although a Median Raphe cyst was not suspected, surgery was required to remove the lesion due to the lack of response to antibiotic therapy. Surgical resection is the treatment of choice for median raphe cysts and may be performed at the patient and physicians’ discretion on a case by case basis [5].

Histologically, there are four different subtypes found in median raphe cyst: urethral, epidermal, glandular and mixed [4]. The cyst lining present in our case was mixed, comprised of urothelium-like epithelium with scattered mucinous goblet cells and squamous cell epithelium. AB/PAS and mucicarmine stains with appropriate controls were used to highlight the mucinous cells.

Differential diagnoses may include apocrine cystadenoma, epidermal inclusion cyst, and pilonidal disease. Histopathological findings and history are pertinent to obtaining the diagnosis.

References