Case Report

GIANT SCROTAL LEIOMYOMA: REPORT OF A RARE CASE AND REVIEW OF LITERATURE.

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ABSTRACT

Background: Leiomyoma is a benign tumour of smooth muscles that can arise almost anywhere in the body; however, scrotal Leiomyomais a rare entity. It may originate from dartos muscle, epididymis or spermatic cord. The neoplasm has an insidious course, and clinically it may mimic many intrascrotal tumours, including testicular cancers, resulting in diagnostic challenges.
Case report: A 20-year-old student evaluated for a painless right testicular swelling of 1year duration, suspicious of right testicular cancer. He had right radical orchidectomy done, but the histology result of the specimen came out to be para-testicular leiomyoma compressing and causing total atrophy of the right testis, the specimen measured 10x7x6cm and weighed 600g. Post-operative condition and follow up were satisfactory.
Conclusion: Scrotal leiomyoma is rare and when it presents as huge mass can cause challenges in diagnosis and treatment, particularly in developing countries. Close follow-up and molecular testing are important in detecting renal cell carcinoma in case of hereditary autosomal dominant disorders that may coexist with this tumour.
Keywords: Paratestisticular, Giant, Dartos muscle, Leiomyoma.

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INTRODUCTION

Smooth muscle tumours of the male external genitalia are uncommon and have received little interest in the literature.[1] Leiomyomas are benign mesenchymal neoplasms, the great majority of which are located in the uterus closely followed by renal capsule. Rare cases arise from the scrotum, pelvis, urinary bladder, and spermatic cord.[2] To a large extent the distribution of the rare tumours parallels the distribution of smooth muscle tissue in the body. The neoplasm tends to be located more commonly in the genitor-urinary and gastrointestinal tracts and less commonly the skin.
We present a rare case of a huge scrotal leiomyoma that can be confused with other intrascrotal tumours, particularly in developing countries with underprivileged health facilities. This study was approved by the ethics committee of the hospital and written informed consent was obtained from the patient.

Case report:
A 20-year-old undergraduate male presented with painless right testicular swelling, which he noticed one year prior to presentation. The swelling was insidious in onset and had no associated pain, dragging sensation, history of itching or trauma to the perineum. There was no history of cough orweight loss. No history was suggestive of urinary tract infections. Essentialfindings in the genital examination were right hemiscrotal mass involving the right testisand no ulceration of the overlying skin. The mass was not tender or warm to touch, and hard in consistency. The left testis and bilateral spermatic cords were normal. The rest of the abdomen and other systems examination was also normal. Clinical diagnosis of query Right testicular tumour to rule out testicular Tuberculosis was made. Mantoux test, full blood count and Erythrocytes Sedimentation Rate (ESR) were not in keeping with Tuberculosis. The patient was then counselled, prepared and had right Radical orchidectomy done as shown in figure 1A. He was seen 3 months postoperative during follow up, the operation site was examined, and he was doing well [figure 1B].

Discussion
Smooth muscle tumours of scrotum were first described by Forsters in 1858 and are reported to be extremely rare.[4]The neoplasm is a benign tumour derived from smooth muscle,[5] it can originate from any anatomic location of smooth muscle in the body.[6,7,8]
The tumours are grouped into cutaneous types comprising of leiomyomata of the erector muscle of skin, dartos muscle of scrotum, labia majora and the nipple. The second class is leiomyomata derived from muscularis of subcutaneous blood vessels (angiomyomata), and the last group is deep soft tissue leiomyomata.[3]The histology of the

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orchidectomy specimen of our index case was found to be Leiomyoma arising from the dartos muscle of the scrotum, which corresponds with the cutaneous leiomyomata. In the same case, there were no testicular parenchymal tissues seen grossly or histologically other than occasional atrophic cells and fibrosis - these features suggest total atrophy of the testis due to long-standing compressive effect by this large tumour.

Scrotal leiomyomata are rare, and only few cases have been reported involving the spermatic cord, epididymis and dartos muscles in the English literature;[9,7,8] however, those of dartos muscle are the rarest.[2,9,10,6,11,5,12,13,14,15] Based on PubMed literature searched, few cases of scrotal dartos muscle leiomyomata were reported.

Leiomyoma most often occurred in older men within the age range of forth to eight decades with mean age of 63 years,[6,7] although a case of dartoid leiomyoma in a 13-month-old infant has been reported.[9] The mean maximum dimension of the neoplasm was 2.5cm with a range of 0.6-7cm.[7,16] In our case, the patient was 20 years old, and the lesion measured 10x7x6cm and weighed 600g.The clinical presentation of dartos muscle leiomyoma is characteristically slow-growing mass and often painless because it pushes the nerves in the scrotum outward rather than compressing them[6].Most patients present within the average age of 8 years between recognition of the tumour and its surgical removal[4]. In our report, the patient presented within 12 months despite its large size. However, when it is large as in the index case, it can lead to erroneous diagnosis and orchidectomy if not well investigated because of suspicion of testicular malignancy especially in young patient coupled with poor health facilities in most of our centres.

Histologically, typical leiomyomata are composed of whorled, anastomosing fascicles of uniform fusiform and spindle smooth muscle cells.[3] The spindle-shaped cells have indistinct borders and abundant fibrillar, eosinophilic cytoplasm. The nuclei are elongated with blunt or tapered ends and have finely dispersed chromatin and small nucleoli[3] (figures3). Mitotic figures are usually infrequent. Immunohistochemistry shows smooth muscle cells that react with antibodies to alpha-smooth muscle actin(as in the index case figures3-E and F), desmin, and caldesmon.[3] There is immunoreactivity with vimentin, but the intensity of staining and the proportion of cells that stain are less than with the muscle-specificantibodies. Epithelial membrane antigen (EMA) isusually negative.[3]

Cytogenetic studies have shown a non-random inactivation of the X chromosome demonstratedby glucose-6-phosphate dehydrogenase isoformexpression in leiomyoma[3]and that indicates proliferation of a single clone ofsmooth muscle cells. Again most cutaneous leiomyomata are sporadic, but yet some occur in the setting of an underlying hereditary autosomal dominant tumour predisposition syndrome that is associated with increased risk for development of an aggressive form of renal cell carcinoma.[2] This suggests that close follow-up of patients and germline molecular testing are very important in early detection of recurrence, malignant transformation and above all possibility of renal cell carcinoma.The treatment of choice for leiomyoma generally is surgical excision which is curative.[13] Irradiation is contraindicated because it can induce malignant transformation.[13] Recurrence is a potentially ominous sign that requires follow up and thorough investigation to rule out malignancy.[5,13]

Conclusion: Dartos muscle leiomyoma is a rare neoplasm, though it is a benign tumour, some may crop up in the setting of underlying hereditary autosomal dominant disorders with the risk of development of renal cell carcinoma. Close follow-up of patients is important in early detection of recurrence and malignant transformation while germline molecular testing can confirm the possibility of development of renal cell carcinoma.This case report highlights the clinicopathological characteristics of scrotal dartos muscle leiomyoma in order to increase our awareness and thus avoid the possibility of erroneous diagnosis and treatment.

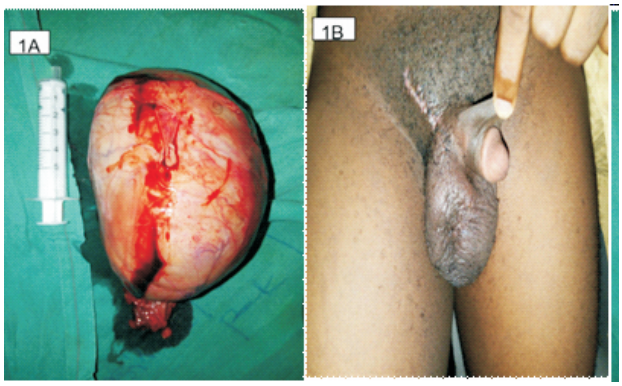

Figure1: Right orchidectomy specimen [figure1A], and postoperative picture during follow-up [figure 1B]

Figure 2: Gross specimen preserved in formalin, showing an ovoid encapsulated grey-white mass that weighed 600g and measured 10x7x6cm, (Figures2A and 2B). Cut surfaces revealed multiple well-circumscribed solid grey white masses, the largest measured 6x5cm(figures 2C and 2D), with whorl pattern. **There is no testicular parenchymal tissue discernable, probably due to the compressive effect of the leiomyoma, leading to testicular atrophy**.

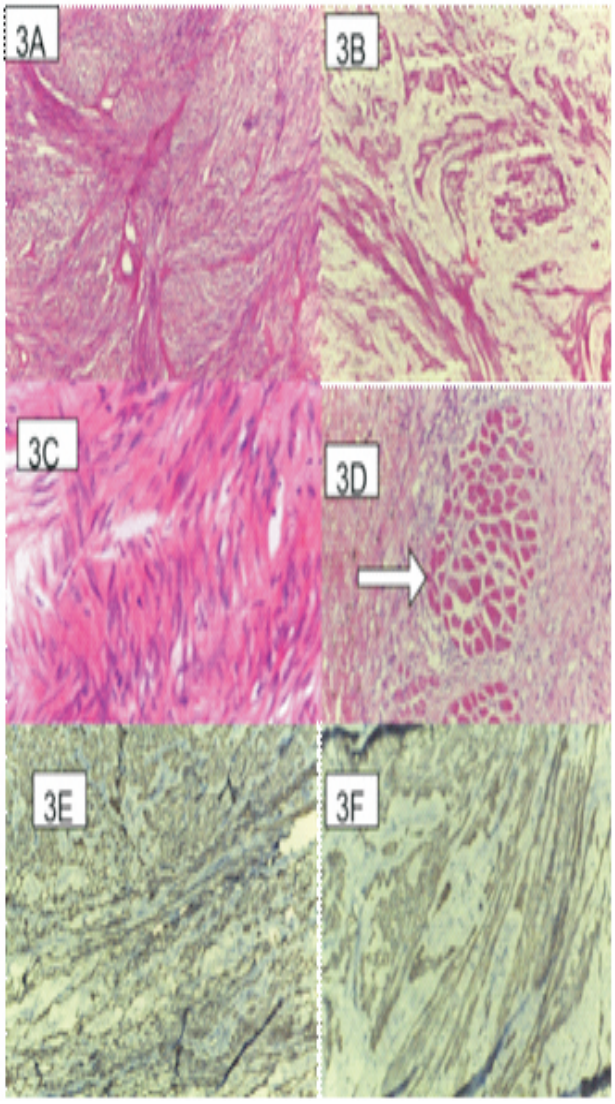

Figure 3: Photomicrographs: The sections show benign mesenchymal neoplasm composed of interlacing bundles of smooth muscle cells (3**A**) that have ovoid to spindle nuclei with tapered ends and moderate fibrillary eosinophilic cytoplasm (3**C**). Section 3**B** shows focal areas of myxoid change. The section from 3**D** shows bundles of dartos muscle (arrow) where the tumour originated. In the photomicrographs above, the testicular parenchyma was effaced by the tumour, and only occasionalatrophic cells were seen along with fibrosis. **Immunohistochemistry** sections were displayed in figures **3E** and **3F**, with positive smooth muscle actin (H and E X100, X100, X200, X100 for 3**A, 3B, 3C** and **3D**, respectively. Immuno: 3**E-** X100 and 3**F**-X200).

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