Epithelioid adenomatoid tumour of epididymis - A rare variant

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Abstract
Adenomatoid tumour is common in the genital organs of both males and females. In the male, epididymis is the most common location. The epithelioid variant of this lesion is very rare and only a few cases have been reported in the English literature. We hereby report a case of epithelioid adenomatoid tumour in the epididymis of a 43 year old patient and forward the typical histopathological microphotographs of the lesion.

Keywords: Adenomatoid tumour, Epithelioid adenomatoid tumour, Epididymis.

Introduction
Adenomatoid tumor is a benign proliferation of mesothelial origin that accounts for 30% of all paratesticular neoplasms and is the second most common tumour after lipoma in this location.[1] Epididymis is the most common location in male genital organs especially in the lower pole. It can also occur in testicular tunica, spermatic cord, ejaculatory ducts, prostate, and suprarenal recess. Rare intra testicular occurrence has also been reported. In women, it is seen in the uterus, fallopian tubes, and less frequently in ovary. It has been reported in extra genital organs also like liver,[2] adrenal,[3] pleura[4] and other organs.

The classic histological appearance is the presence of pseudovascular spaces of varying sizes lined by cuboidal/flattened cells resembling a hemangioma. Epithelioid morphology of this tumour is very rare. Only a few cases have been reported in the English literature. And microphotographs of this variant is rarely depicted in the text books and reported articles. Hence, we are reporting our case of epithelioid variant of adenomatoid tumour in the epididymis of a 43 year old patient with microphotographs of the same.

Case Report
43 year old patient presented with painless swelling in the lower pole of left epididymis since last 2months (Fig. 1a). Clinical diagnosis was epididymal cyst/tumour. Excision of the lesion was done and we received the specimen as a linear piece of epididymal tissue with an attached nodular lesion towards one pole. Nodule measured 2x1.5x1cm. Portion of epididymis measured 2.5 cms in length. Cut section of the nodule showed a well circumscribed pale white homogenous lesion measuring 2 cm in greatest dimension. [Fig. 1b]

Microscopy of the lesion on H&E staining showed a well circumscribed, unencapsulated neoplasm composed of cells arranged in cords, tubules and sheets [Fig. 2]. In between slit like spaces of varying sizes were seen surrounded by similar type of cells [(Fig. 3]. The Cells were large polygonal with abundant eosinophilic cytoplasm, round to oval nuclei many showing prominent nucleoli [Fig. 4a & b]. Small intra cytoplasmic vacuoles were seen, some coalescing together [Fig. 5a]. No cellular or nuclear pleomorphism noted. Mitosis was scanty. Stroma was fibrous, intermixed with bands of smooth muscle fibres [Fig. 5b]. In areas, dense lymphocytic cell aggregates were noted [Fig. 6]. A few thick walled blood vessels noted. Periphery showed epididymal tissue.

The provisional diagnosis from the routine hematoxylin & eosin stained sections was epithelioid adenomatoid tumour because of the epithelioid nature of the cells surrounding tiny cystic spaces. But the tumor morphology resembled a sex cord stromal cell tumour especially leydig cell tumour. Also, the possibility of an Ectopic adrenocortical tissue could not be excluded. Since sertoli cell nodules, leydig cell nodules and ectopic adrenocortical tissue may be rarely seen in paratesticular region including epididymis, these were kept as differentials. Hence immunohistochemical study was done to confirm the diagnosis. The markers studied were antibodies to Pan CK, Calretinin, Inhibin and S100. The tumour showed strong diffuse positivity for Pan CK and Calretinin confirming the differentiation towards mesothelial cells. Inhibin and S100 were negative in the tumour cells [Fig. 7]. Ki 67 immunostain showed low proliferation index.

Discussion
Adenomatoid tumour is a benign neoplasm of mesothelial origin. In older literature it was described in various names like adenoma, lymphangio-endothelioma, benign adenomatous tumour, lymphangioblastoma and a variety of other names because of the obscurity of the histogenesis of this tumour at that time. In 1945, Golden and Ash first proposed the non-committal term "adenomatoid" tumour.[5] The authors thought that the the proposed name has the advantage of being morphologically

correct and genetically neutral. Masson et al in 1942[6] and Evans in 1943[7] suggested a mesothelial origin for these tumors.

It is seen in a wide range of ages and with a peak incidence between 20 and 50 years. Size varies from a few millimeters to 5 cm. The present case is of a 43 year old patient with a 2 cm size lesion. The most common location in male is the lower pole of epididymis as is the present case. Our cases showed the classical features of slit like spaces bordered by epithelial like cells with intracytoplasmic vacuoles, lymphoid infiltrate and admixed smooth muscle bundles. But instead of the classic cuboidal/flattened lining cells, this case showed cells with an epithelioid morphology composed of large polygonal cells with abundant eosinophilic cytoplasm – the Epithelioid variant of the tumour which was confirmed by appropriate immunohistochemical marker study. In a report by AR Sangoi et al who studied both male and female genital tract adenomatoid tumours, lymphoid aggregates, either admixed with tumor cells or around the periphery, was seen in all male genital tract adenomatoid tumors ranging from focal to moderate in extent. Among the female genital tract sites, lymphoid aggregates were infrequent and when present, were focal.[8]

Mario Alvarez Maestro et al[9] studied 9 cases of adenomatiod tumor of male genital organs and found the classical glandular morphology in 8 cases and alveolar pattern in 1 case. None of their cases had epithelioid morphology. Lymphoid proliferation was found in all of their cases. We also found lymphoid infiltrate in the present case forming follicles within the tumour. Many reports showed varying amount of smooth muscle fibres admixed with the tumour which was present in this case also.

Various studies have been conducted with different immunohistochemical markers for mesothelial cells like Pan CK, Calretinin, WT1, D2-40, CK5/6, calponin and others. AR Sangoi et al found calretinin, D2-40 and WT1 more sensitive than the other mesothelial markers.[8] In the present case also the tumour cells showed strong diffuse positivity for Calretinin and pan CK confirming the mesotheliial nature of the neoplasm.

Epithelioid variant of adenomatoid tumour does not have an aggressive behaviour or malignant potential. The significance as far as pathologists are concerned is that it should not be misdiagnosed as some other tumour because of the unusual morphology. Local excision is the treatment of choice since the tumour has a benign behaviour.

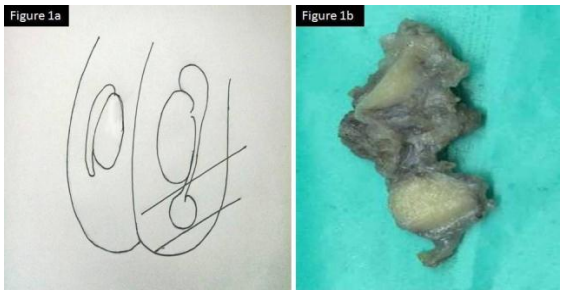

Fig. 1a: Lesion in the lower pole of Lt. Epididymis Drawing by clinician
Fig. 1b: Cut section of the lesion- Circumscribed grey white lesion with part of epididymis

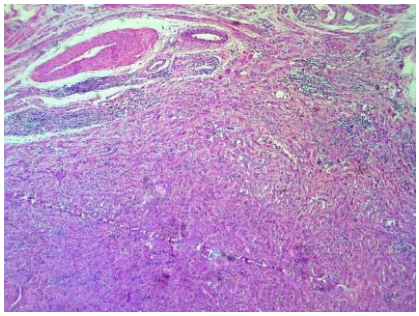

Fig. 2: Unencapsulated, circumscribed tumour (H&E x40)

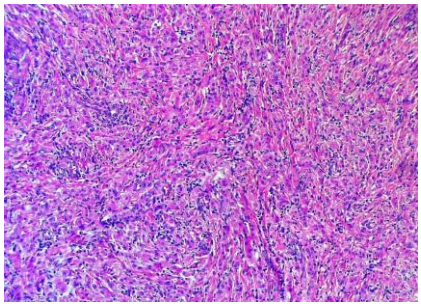

Fig. 3: Tumour tissue with epithelioid cells and pseudovascular spaces (H&E x 100)

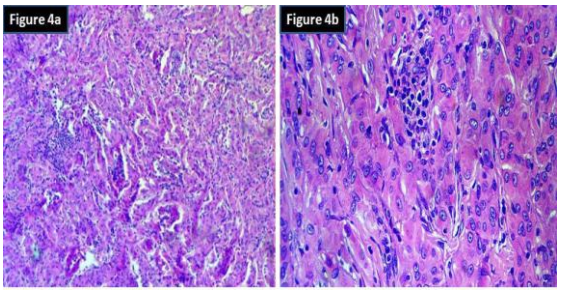

Fig. 4a: Slit like spaces within the tumour -Higher magnification (H&E x 400)
Fig. 4b: Epithelioid cells in the tumour -Higher magnification (H&E x 400)

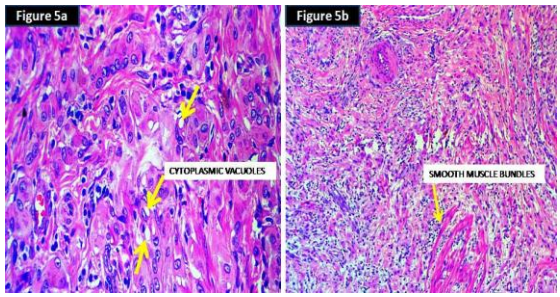

Fig. 5a: Intra cytoplasmic vacuoles (H&E x 400)
Fig. 5b: Smooth muscle bundles in the tumour (H&E x 400)

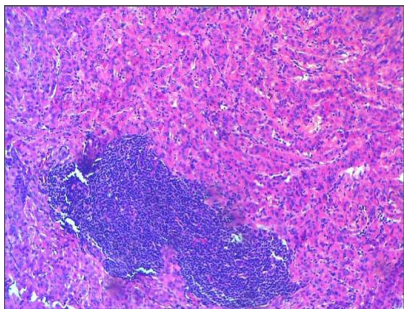

Fig. 6: Lymphoid aggregate in the tumour (H&E x 400)

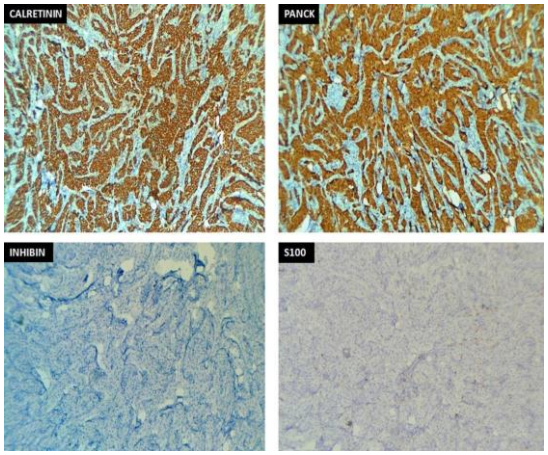

Fig. 7: Immunohistochemical stains (x100)

Conclusion

Epitheliod adenomatoid tumour, a rare variant of adenomatoid tumour is reported in the epididymis of a 43 year old patient with microphotographs because of the rarity of this variant. The tumour has a benign behaviour and hence surgical excision is curative. The tumour should not be misdiagnosed as a carcinoma because of the epithelioid morphology.

Consent: Written informed consent was obtained from the patient for publication of this case report and any accompanying images.

Competing Interests: The author(s) declare that they have no financial or non financial competing interests.

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