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EPITHELIOID HEMANGIOENDOTHELIOMA OCCURING IN A CRYPTORCHID TESTIS: A RARE TUMOR AT A RARE SITE

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ABSTRACT

Epithelioid hemangioendothelioma (EHE) is a rare low-grade sarcoma of endothelial origin, and was originally described by Weiss and Enzinger. The tumor has been described to occur at various soft tissue sites and even in visceral organs. The cryptorchid testis is a known site for malignant neoplasms but the commonest tumors arising here are carcinomas, most commonly germ cell tumors. We herein report a case of EHE in a cryptorchid testis of a 55 year old male which showed positivity for the vascular markers, CD 31 and CD 34.

Keywords : cryptorchid testis, epithelioid hemangioendothelioma, positivity for CD 31, CD 34

INTRODUCTION

Epithelioid hemangioendothelioma (EHE) is a rare malignant tumor of vascular origin. The tumor usually occurs in superficial or deep soft tissue and is described as a single reddish-white mass, slightly painful on palpation. In many cases the tumor is related to vessels, especially to veins. Cryptorchid testes are usually found in the inguinal canal (80 % of cases); they are prone to trauma, torsion, inguinal hernia (10-20 % of cases), and sterility, and are associated with 5 - 50 times increased risk of testicular carcinoma, usually seminoma and other germ cell tumors. Mesenchymal tumors are rare and to the best of our knowledge, EHE has not been reported in the cryptorchid testis till date. We report a case of a 55 year male with a provisional diagnosis of left inguinal hernia and left undescended testis. Histopathology of the testis revealed EHE.

CASE REPORT

A 55 year old male patient presented to the outpatient department of our hospital with a history of swelling and pain in the left groin for the past eight years. The swelling extended till the scrotum and disappeared on lying down. An ultrasound of the swelling revealed an oval-shaped hypoechoic structure in the left inguinal canal which was radiologically suspected to be an undescended or cryptorchid testis. A provisional diagnosis of left inguinal hernia with left undescended testis was made and the patient was taken up for left hernioplasty with orchidectomy. We received the orchidectomy specimen measuring 3 x 2.5 x 0.5 cms, along with a sac measuring 0.5 cm, and a cord 8 cm in length. Cut surface of the orchidectomy specimen showed an atrophic testis with reddish-brown areas. String sign was positive in the testis. Microscopic examination of the same showed hyalinised and atrophic testicular tubules surrounded by nests of rounded and slightly spindled eosinophilic endothelial cells with round nuclei and prominent cytoplasmic vacuolization (with few containing erythrocytes in them). Small capillary lumina at the edge of the tumor were also discernible. (Figure 1 A-C) An occasional mitotic figure and mild atypia were also noted. Immunohistochemistry with CD 34 and CD 31 showed strong positivity for the endothelial cells (Figure 1D).

Microscopic examination of the sac revealed fibrocollagenous tissue with inflammatory cellular infiltrate while the cord showed normal cord structures. A final diagnosis of EHE in an atrophic cryptorchid testis was made.

DISCUSSION

EHE was first described by Weiss and Enzinger as a tumor composed of short strands or solid nests of rounded to slightly spindled endothelial cells.[1] Distinct vascular channels are uncommonly seen except in the more peripheral portions of the tumor. The tumor cells instead form small intracellular lumina (few containing red blood cells) which are seen as clear spaces (vacuoles) that distort or "blister" the cell.[1] The stroma can be myxoid or hyaline. Usually the tumor is bland with virtually no mitotic activity although in about one-fourth of cases, the tumor can show high-grade features like significant atypia, prominent mitotic activity (> 1/10 HPF) or necrosis.[1] EHE has been reported in several sites including in the liver,[2,3] lungs,[4] head and neck, bone,[5,6] heart,[2] spermatic cord, paraovarian region and spine.[7] The tumor is usually positive for CD 31 and CD 34 immunohistochemical stains thereby reiterating its origin from blood vessels. At the same time, the poor expression of Ki67 and weak positivity for VEGF (a growth factor specific for vascular endothelial cells) is indicative of the less aggressive nature of the lesion.

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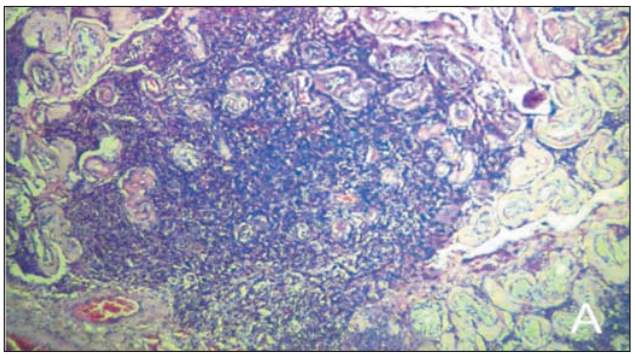
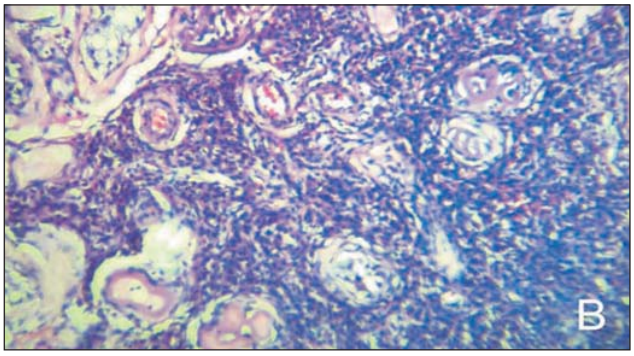
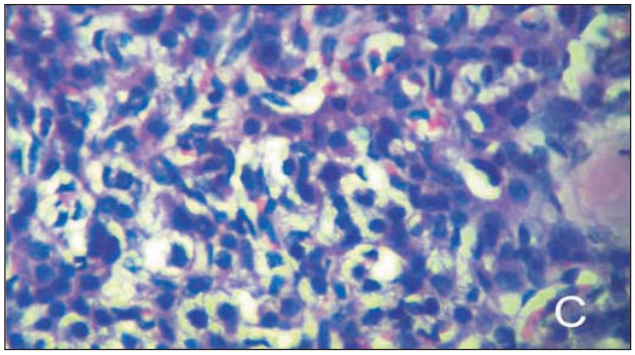


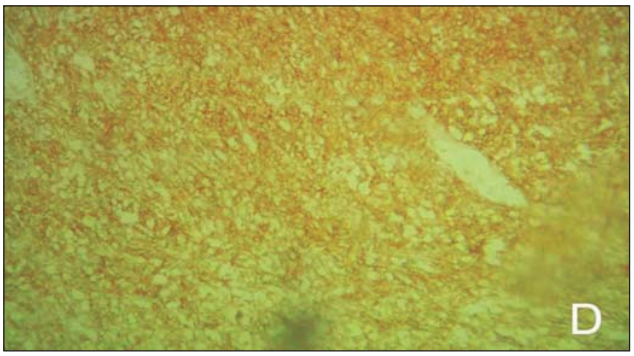
A. Scanner view (H&E x 40) showing hyalinised testicular tubules amidst a cellular neoplasm.



B. Low power view (H&E x 100) showing a highly cellular neoplasm composed of round to oval cells with eosinophilic cytoplasm; few capillaries with intraluminal RBCs can be seen at the periphery of the tumor.



C. High power view (H&E x 400) showing rounded endothelial cells with hyperchromatic nuclei, prominent eosinophilic cytoplasm and characteristic intracytoplasmic vacuolations or "lumina" with few of them containing RBCs.



D. Scanner view (H&E x 40) of the tumor showing intense CD 34 cytoplasmic positivity of the tumor cells.

The differential diagnoses for EHE include metastatic adenocarcinoma or melanoma and also many sarcomas that may adopt an epithelioid appearance. In uncertain cases, immunohistochemistry as well as electron microscopy may be helpful in the differentiation.[8] The overall prognosis of EHE, although not extensively studied, seems quite favorable.

Literature search till date has not got a single report of EHE in a cryptorchid testis. Patients with cryptorchid or undescended testis usually have a substantially increased relative risk of testicular malignancies particularly when they present clinically as inguinal hernias,[9] as in our case. The excess risk associated with undescended testis is eliminated in men who had an orchidopexy before the age of 10 years. In a study of 137 patients with germ cell tumors of the testis associated with cryptorchid testis, the histologic type of the tumor was pure seminoma in 56 patients, embryonal carcinoma in 41, teratocarcinoma in 37 and pure choriocarcinoma in 3 patients.[10] Uncorrected cryptorchidism carries a higher risk for seminoma while corrected ones have a greater risk of non-seminomatous tumors.

Cryptorchid patients with testes that descended late should be observed periodically, especially after the 20-year latent period, for early detection of cancer. In our patient, the undescended testis had been noted only at the age of 55 years which confirms the fact that later the testicular descent, higher is the chance of developing malignancy. This case is being reported for its rarity and to create the awareness that undescended testis are sites for mesenchymal tumors also and not just germ cell tumors or other carcinomas.

REFERENCES

1. Weiss SW, Enzinger FM. Epithelioid hemangioendothelioma: a vascular tumor often mistaken for a carcinoma. Cancer 1982;50:970-81.
2. Hurley TR, Whisler WW, Clasen RA, Smith MC, Bleck TP, Doolas A, Dampier MF. Recurrent intracranial epithelioid hemangioendothelioma associated with multicentric disease of the liver and heart: Case report. Neurosurgery 1994;35:148-51.
3. Ishak KG, Sesterhenn IA, Goodman ZD, Rabin L, Stromeyer FW. Epithelioid hemangioendothelioma of the liver: a clinicopathologic and follow-up study of 32 cases. Hum Pathol 1984;15:839-52.
4. Dail DH, Liebow AA, Gmelich JT, Friedman PJ, Miyai K, Myer W, et al. Intravascular, bronchiolar and alveolar tumor of lung (IVBAT): An analysis of 20 cases of a peculiar sclerosing endothelial tumor. Cancer 1983;51:452-64.
5. Abrahams TG, Bula W, Jones M. Epithelioid hemangioendothelioma of bone: a report of two cases and review of literature. Skeletal Radiol 1992;21:509-13.
6. Kleer CG, Unni KK, Mcleod RA. Epithelioid hemangioendothelioma of bone. Am J Surg Pathol 1996;20:1301-11.
7. McCluggage WG, Dolan S, Cameron CHS, Russelt CFJ. Epithelioid hemangioendothelioma of the spermatic cord. Int J Surg Pathol 2000;8:75-8.
8. Pillay K, Chetty R. Epithelioid hemangioendothelioma of the paraovarian region. Journal of Gynecologic Surgery 2002;18:33-6.
9. Pinczowski D, Mclaughlin JK, Lackgen G, Adami HO, Persson I. Occurrence of testicular cancer in patients operated on for cryptorchidism and inguinal hernia. J Urol 1991;146:1291-4.
10. Batata MA, Chu FC, Hilaris BS, Whitmore WF, Golbey RB. Testicular cancer in cryptorchids. Cancer 1982;49:1023-30.