INTRODUCTION

Adenocarcinoma of rete testis is a rare tumor with only 60 cases reported till date and with an average survival of only eight months. The dynamics of the disease have been a topic of much deliberation as its rarity renders it difficult to be understood. The revised criteria by Nochomovitz are used for diagnosis. The commonest presentation is in the form of a mass in the scrotum which is usually painless. We present a case of adenocarcinoma of rete testis presenting as a hydrocele.

CASE REPORT

A 58-year-old male had presented to our hospital with complaints of pain and swelling in the left hemiscrotum since seven days with continuous fever. The patient had a left eversion of sac two weeks back. He was febrile, had tachycardia (pulse: 110/min), and was well nourished. He had a swelling in the left hemiscrotum of size 9 × 8 cm with tenderness. Fluctuation test of the left hemiscrotum was positive but transillumination test was negative. Rest of the abdominal and other systemic examination was unremarkable. His complete blood count showed an increase in the total leukocyte counts (TLC) to 12,350/cubic mm with 80% polymorphonuclear leukocytes. With a provisional diagnosis of postoperative pyocele, exploration was planned. On exploration, there was a purulent discharge from the sac. There was a hard mass at one pole of the testis. Hence, a high inguinal orchidectomy was done, and postoperatively, the patient made an uneventful recovery. The gross examination of the specimen showed a gray-white, ill-defined, 2.5 × 1.5 cm sized solid tumor located in the rete testis. Histologically, the tumor was a well-differentiated adenocarcinoma with papillae, arising from the rete testis. Intraductal spread into seminiferous tubules was noted. There was no extension of tumor to the testicular parenchyma or tunica [Figure 1]. Histopathologically, a diagnosis of adenocarcinoma of rete testis was confirmed. Retrospectively, in the light of...
histopathological diagnosis, a search from primary was initiated. His rectal examination demonstrated a normal prostate. Chest X-ray and abdominal ultrasound were also normal. Laboratory investigations like alkaline phosphatase, serum alpha fetoprotein (AFT), and prostate-specific antigen were also normal. Computerized tomography of the abdomen could not be performed due to economic constraints. With this, a diagnosis of a primary nature of adenocarcinoma was confirmed. The patient opted for palliative treatment and was discharged with advice to follow-up.

DISCUSSION

Adenocarcinoma of rete testis is a rare tumor with only 60 cases reported globally till date.[1] Due to the rarity and poor prognosis of the tumor, the natural course and outcomes of treatment have been poorly understood. Most of the tumors are incidentally discovered. It may present as a painless solid or cystic intrascrotal mass (94%), and the diagnosis is clinched by histopathology. Twenty-seven percent patients may present with scrotal pain and 19% patients may present with signs and symptoms due to metastasis.[2] Sometimes the tumor presents in association with hydrocele (25%),[1] hematocoele,[3] epididymitis, inguinal hernia, or undescended testis.[1] In our case, the tumor had presented as a hydrocele.

The tumor can present in any age group, but is most commonly seen in elderly males over 60 years of age. The reported five-year survival of the tumor is only 13%.[3] Mostly the adenocarcinoma is a primary tumor, but it may be secondary and the incidence varies between 0.02% and 2.5%. Metastasis from prostate is most common, followed by lung.[3]

The tumor is diagnosed histologically by revised Nochomovitz criteria which include: (1) location of tumor around hilum, (2) no involvement of the tunica, (3) absence of histologically similar extra-scrotal tumor that plausibly could be the primary, and (4) immunohistochemical exclusion of other possibilities, particularly malignant mesothelioma and papillary serous carcinoma.[4] Primary adenocarcinoma on gross appears as a poorly circumscribed, solid, firm, and rubbery nodule with a cystic component occasionally. It is located within the tunica albuginea at testicular mediastinum. Microscopically, it has either a glandular or papillary pattern with intraductal spread into seminiferous tubules being characteristic, which also helps in distinguishing the tumor from mesothelioma.[5] In our case, most of the criteria for diagnosis was fulfilled except immunohistochemical exclusion of other tumors like malignant mesothelioma and papillary serous carcinoma. Visualization of a transition from normal to neoplastic rete epithelium is also a useful diagnostic clue if present, but large masses often obliterate the normal anatomy such that a transition is not evident.[6] The differential diagnosis includes malignant mesothelioma, certain ovarian-type tumors, metastatic adenocarcinoma, epididymal carcinoma, and malignant Sertoli cell tumor.[7] Mullerian-type serous carcinomas have a predominantly cystic component and have plainer cytology as compared to rete adenocarcinoma.[7] Metastatic adenocarcinoma can be differentiated from primary adenocarcinoma by clues as follows: (1) It is very commonly bilateral and/or multifocal, (2) demonstrates frequent vascular-lymphatic invasion, (3) has an interstitial pattern of growth (in between rather than from the rete epithelium), and (4) usually occurs in patients with a history of a primary carcinoma, although sometimes the history is remote.

A search for reliable tumor markers for the diagnosis of adenocarcinoma of rete testis has been in vain. A few cases have been examined immunohistochemically and are positive for cytokeratins and negative for β-hCG (beta human chorionic gonadotropin) and AFP. Positivity for mesothelioma-related markers (positive calretinin, WT-1, CK 5/6, and negative CEA, Leu-M1, Ber EP4, and B72.3) is helpful in confirming the diagnosis of mesothelioma.[6] Likewise, Ca-25 and WT-1 are important in confirming a Mullerian-type derivation.[6]

Adenocarcinoma of rete testis is an aggressive malignancy with poor prognosis. The treatment is primarily surgical with high inguinal orchidectomy. Radiotherapy, chemotherapy, and alternative surgical methods have been tried. Survival is dismal, with an average survival of only eight months. Careful follow-up is necessary.

REFERENCES


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