Paratesticular Malignant Mesothelioma - A Rare Case Presentation

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Abstract Paratesticular malignant mesotheliomas are rare tumors with 223 cases described so far. It is a highly aggressive tumour. We report a case of a malignant mesothelioma arising from the tunica vaginalis of the testis which was highly aggressive in its course.

Keywords Paratesticular malignant mesothelioma · Calretinin · Hydrocele · Orchidectomy

Introduction

Paratesticular mesotheliomas are rare tumors. The sole plausible causative factor so far ascertained in the pathogenesis of these tumors is asbestos, which however is found in only around 30–40 % of such cases [1]. The age range of affected individuals is wide, mostly adults and the elderly, but also includes young people and children. We present a case of a malignant mesothelioma in a young male which presented as testicular torsion and came as a histological surprise.

Case Report

An 18-year-old man was admitted to the ward of surgical Unit IV at Coimbatore Medical College in January 2009 with complaints of acute lower abdominal pain, acute scrotal pain with scrotal swelling, with no other specific complaints. There was no history of fever or trauma preceding

the onset of scrotal pain or swelling. Physical examination of the abdomen revealed tenderness in the left iliac fossa. Examination of the scrotum showed a swelling arising from the left testis which was warm and tender. Cord structures appeared to be thickened. The ultrasound scan of the abdomen was normal. Doppler ultrasound of the scrotum revealed left testicular torsion with loss of vascularity to the left testis.

It was decided to proceed with an emergency scrotal exploration on the basis of the ultrasound scrotum report. On exploration, we found a mass arising from the left testis. Left orchidectomy was done (Fig. 1). Histopathology of the excised left testis revealed a paratesticular malignant mesothelioma arising from the tunica vaginalis of the left testis with underlying epididymo-orchitis (Fig. 2). Immunohistochemical analysis showed intense expression of cytokeratins, vimentin, and calretinin.

Postoperatively, a thorough evaluation of the abdomen and thorax was done by means of computed tomography, which were found to be normal. The patient was started on chemotherapy with adriamycin and paclitaxel-based regimen. He developed rib, skeletal, and skull metastasis within 10 months of diagnosis (Figs. 3 and 4). He received external radiotherapy for the skull and skeletal metastasis. The patient died after 14 months of primary diagnosis.

Discussion

Malignant mesothelioma is an uncommon tumor arising in body cavities lined by mesothelium. The majority of these tumors are found in the pleura, peritoneum, and less frequently pericardium. As the tunica vaginalis is a layer of reflected peritoneum, mesothelioma can occur in the scrotal sac. The nonspecific symptoms, broad age distribution, and

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Fig. 1 Specimen of excised left testis

lack of tumor markers make preoperative diagnosis of malignant mesothelioma of the tunica vaginalis difficult and indeed most of the reported cases are diagnosed at histology. Although ultrasound is considered the imaging modality of choice in evaluation of scrotal tumors, to date, reports on the ultrasound features of mesothelioma arising from tunica vaginalis are limited [2].

The most common presenting symptom is either hydrocele of unknown origin or intrascrotal mass. When hydrocele is the presenting symptom, these tumors are often clinically overlooked and the diagnosis is delayed [3]. Most paratesticular mesotheliomas arise in the tunica vaginalis, but primary tumors of the spermatic cord and epididymis are also on record. Tumors arising from the peritoneal mesothelium of a hernia sac are excluded from this group. The correct diagnosis is almost always made after histologic examination of the operative specimen. Immunohistochemistry and electron microscopy are always helpful and sometimes necessary tools for diagnosis. So far a very few cases have been identified or

Fig. 2 Histopathology result showing highly cellular smears composed of large, plump cells distributed mainly in cohesive clusters. Cellular dissociation with single cells and smaller groups was present. Neoplastic cells showed a moderate to abundant amount of cytoplasm and polygonal to slightly elongated morphology

suspected preoperatively on cytologic examination. Three clinicopathologic types of malignant mesotheliomas of the male genital tract are recognized: diffuse tubulopapillary mesothelioma, well-differentiated papillary mesothelioma, and multicystic mesothelioma. The histologic subtypes are almost always pure epithelial or biphasic. The differential diagnosis is mainly with serous papillary tumors arising from Mullerian vestiges, but several diverse primary or secondary tumors also need to be considered.

The first primary cytological diagnosis of a malignant mesothelioma of the tunica vaginalis of the testis was reported by Japko et al. and Gorini et al. They reported two cases of malignant mesothelioma of the tunica vaginalis of the testis, both with a history of asbestos exposure for more than 10 years. One case was a purely epithelial type and the other case was biphasic. The full range of growth patterns of a malignant mesothelioma can be seen by using fine-needle aspiration cytology, including highly differentiated epithelial tumors as well as biphasic, sarcomatous, and anaplastic forms. The most specific cytological pattern is a combination of sheets of cells, cell groups, dispersed polygonal cells with dense cytoplasm, epithelial cells, and spindle-shaped fibroblastic cells. Occasionally, a biphasic pattern is seen with both spindle cell and pure epithelial cell components [4]. Eleven cases of malignant mesotheliomas of testicular tunica vaginalis were reported in the age range of 12–76 years by Jones et al. [5]. Microscopic analysis revealed that 5 of these 11 tumors were epithelial and 6 were biphasic with typical architectural and cytological features of a mesothelioma. Mixtures of papillary, tubular, and solid patterns predominated in the epithelial areas, whereas interlacing fascicles of spindle-shaped cells with scanty stroma characterized the sarcomatous component.

The major difficulty in managing patients with malignant mesothelioma of the tunica vaginalis testis is in determining an accurate preoperative diagnosis. Due to the lack of characteristic symptoms, most of the cases are diagnosed intraoperatively. Those patients who undergo local resection of the hydrocele wall, 35.7 % experience local tumor recurrence, as compared with 10.5 % after scrotal orchidectomy and 11.5 % after inguinal orchidectomy. Therefore, radical orchidectomy should be the first-line therapy. The median survival of the patient is 23 months. The overall recurrence rate (local and disseminated) is 52.5 %. More than 60 % of recurrences develop within the first 2 years of the follow-up. In some cases of disseminated mesothelioma, adjuvant chemotherapy or radiotherapy is given. Although reports on adjuvant treatments were limited, radiotherapy appears to be more effective than chemotherapy. Assessment of prognostic parameters has revealed a significant correlation of patient's age with survival, with a better outcome for younger patients and a worse disease course for patients with primary disseminated disease in univariate analysis. About 38 % patients die of disease progression [6].

Fig. 3 MRI showing skull metastasis

Fig. 4 MRI showing skeletal metastasis

Conclusion

Malignant mesotheliomas of the tunica vaginalis testis rarely occur, but the possibility should be considered for all age groups. Univariate analysis determines that a patient's age and the presence of primary disseminated disease are prognostic parameters related to survival. Due to the invasive potential of this disease and the risk of tumor recurrence, radical orchiectomy and close follow-up are strongly recommended. But the overall prognosis and survival are poor.

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