CASE REPORT

Malignant mesothelioma of the tunica vaginalis

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Introduction: *Malignant mesothelioma involving the tunica vaginalis is an extremely rare tumor.*

Case: *We describe a case of a 39-year-old man who initially presented with discomfort in the left testis and underwent resection of a hydrocele in the left testis. A pathology examination of a frozen section of a lesion on the tunica identified the lesion as mesothelioma, which was confirmed by a second pathology examination. No metastatic disease was found. The patient subsequently underwent a left radical orchiectomy and a partial scrotectomy, and has been disease free for 7 years.*

Discussion: *The first case of malignant mesothelioma involving the tunica vaginalis, a structure embryologically derived from a layer of reflected peritoneum known as the "processus vaginalis," was reported in 1957. Only 73 cases were reported in the literature between 1966 and 1997. In up to 41% of cases, there is a positive history of asbestos exposure. Young age and localized disease are associated with a better prognosis. Radical orchiectomy limits recurrence, which usually occurs within 2 years, but may occur up to 15 years after surgery.*

Key Words: malignant mesothelioma, tunica vaginalis, radical orchiectomy, testicular neoplasm

Introduction

Malignant mesothelioma of the tunica vaginalis is a rare tumor, and only about 80 cases have been reported in the literature up to 2005.[1] Most patients are initially treated conservatively for other potential causes of scrotal swelling. The diagnosis of malignant mesothelioma of the tunica vaginalis is most often made postoperatively by the pathologist, or, on occasion, is suspected after conservative patient management fails.

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Case report

A 39-year-old man presented to our urology clinic in 1999 complaining of discomfort in his left testis. A diagnosis of epididymal orchitis was made, and the

patient was prescribed a course of antibiotics. The epididymal orchitis resolved. Three weeks later, the patient presented with an enlarged left scrotum. Physical examination and a subsequent ultrasound confirmed that the swelling was a hydrocele. The patient's physical examination was otherwise normal. He did not have any past history of any urological or other medical events and had no personal or family history of asbestos exposure. His laboratory levels of alpha-foeto protein and beta human choriogonadotropin were normal.

The patient underwent surgery for a routine hydrocele repair. His left testis was explored via a midline scrotal incision. A lesion that appeared inflamed was identified on the tunica. This lesion was excised and sent for pathology examination of a frozen section. The initial pathology report identified the lesion as a mesothelioma. A hydrocelectomy was performed (rather than a radical orchiectomy, since mesothelioma is rare). The final pathology report confirmed that the tunica lesion was a malignant mesothelioma of a mixed epithelial and sarcomatoid type.

Subsequent computed tomography (CT) scans of the patient's chest, abdomen, and pelvis failed to reveal metastatic disease. Three weeks later, the patient underwent a left radical orchiectomy and a partial scrotectomy. At regular follow-up examinations every 6 months for 7 years, no evidence of recurrent disease was found.

Pathology

Macroscopically, the tumor consisted of a firm fibrotic plaque of the tunica vaginalis, measuring 2.1 cm x 1.4 cm x 0.4 cm. Microscopically, the tumor was biphasic with epithelial and sarcomatous components. There were mixed epithelial cell patterns: tubular, solid, and papillary, with occasional psammoma bodies. The sarcomatous areas showed atypical spindled cells in loose fibrillar arrangements. The tumor was clearly malignant, showing invasion with a desmoplastic stroma, striking cytological atypia, and frequent mitoses. The background was one of atypical mesothelial hyperplasia merging with malignant mesothelioma in-situ of the tunica vaginalis. Electron microscopy confirmed the mesotheliomatous nature of the neoplasm, revealing abundant "spaghetti" microvilli. Figures 1-3.

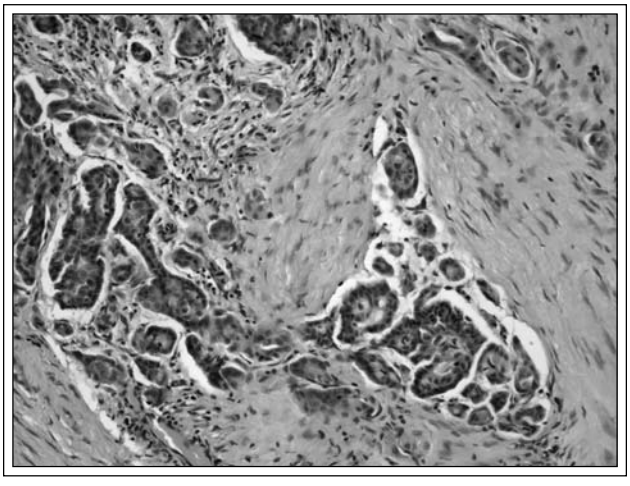

Figure 1. Papillary arrangements of malignant mesothelioma in fibrous stroma (400X).

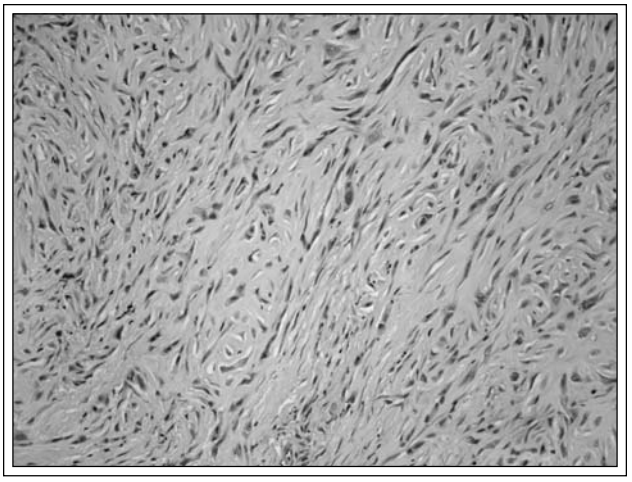

Figure 2. Spindled malignant mesothelial cells in dense stroma form the sarcomatous component (400X).

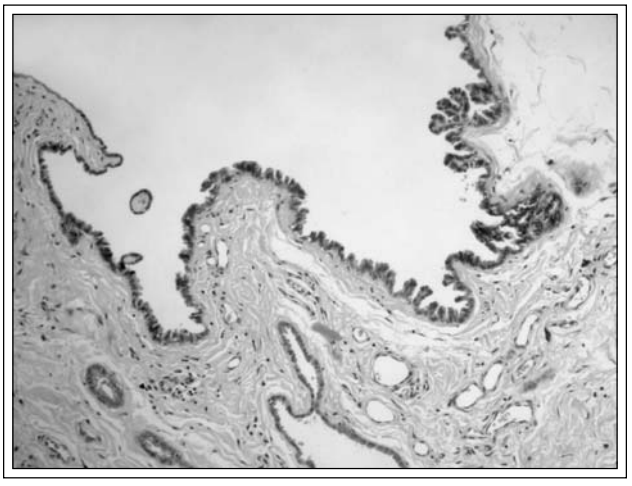

Figure 3. Mesothelium changes from benign cuboidal (left), through hyperplastic area to architecturally and cytologically atypical in situ mesothelioma (right) (200X).

Discussion

Malignant mesothelioma is a rare tumor derived from mesothelial cells. There are approximately 2000 cases of mesothelioma each year in the United States.[2] The tumor most commonly arises in the pleura, but 10% to 20% of cases arise in the peritoneum. Mesotheliomas originating in the tunica vaginalis are extremely rare. More than 80% of mesothelioma cases can be attributed to asbestos exposure. The first documented case of mesothelioma of the pleura occurred in an asbestos worker in England in 1946.[3] Other etiological factors that have also been implicated in the development of mesothelioma include exposure to simian virus 40, radiation, or Thorotrast (a colloidal suspension of radioactive thorium dioxide).[4] At the molecular level, chromosomal abnormalities such as losses on 1p, 3p, 6q, 9q, and monosomy 22 have been reported. These may explain mechanisms for the oncogenesis of mesothioloma.[5] Three subgroups of malignant mesothelioma have been identified. There is a tendency for pleural mesotheliomas to be sarcomatous, for peritoneal mesotheliomas to be epithelial, and for those from serosal surfaces to be biphasic.[6]

The case reported here involves a man with a rare, malignant mesothelial tumor arising from the tunica vaginalis. The tunica vaginalis arises embryologically from a layer of reflected peritoneum known as the "processus vaginalis," which lines the scrotal pouch. Atrophy occurs, except for the portion that surrounds the testis. The first case of malignant mesothelioma of the tunica vaginalis was reported in 1957 by Barbera and Rubino.[7] This type of tumor is rare, and from 1966 to 1997 only 73 cases were reported in the literature.[8] It accounted for only 0.09% of deaths due to mesothelial tumors in the United Kingdom from 1968 to 1991, according to the United Kingdom Health and Safety Executive (HSE) mesothelioma register.[9]

Unlike other malignant mesotheliomas, no clear etiological factor has been established for those arising from the tunica vaginalis. However, a positive history of exposure to asbestos was present in up to 41% of cases according to Jones and colleagues[10] and Plas and colleagues.[8] Our patient did not have any personal or family history of asbestos exposure, and no other etiological factor was identified. Due to the rarity of this tumor type, it will likely to be difficult to identify other potential causative factors associated with it. Reviews of reported cases have identified prognostic factors associated with better survival. Plas and colleagues[8] reported that younger age was associated with a better outcome ($p < .001$) and primary disseminated disease was associated with a worse prognosis ($p < .05$).

Tumors of the tunica vaginalis tend to occur in older men; the peak incidence is in men older than 50 years old. However, 10% of cases occur in men younger than 25 years old.[8] As is the case with malignant mesotheliomas that arise from the pleura and peritoneum, those arising from the tunica vaginalis tend to have an aggressive clinical course. Although most patients present with a localized testicular mass, patients may also present with local invasion of the skin of the scrotum, epididymis, and penis. Patients may also present with distant metastases — most commonly in the retroperitoneal and inguinal lymph nodes, and, occasionally, in the lung and abdominal peritoneum.[9]

There are as yet no established clinical practice guidelines for patient treatment and follow-up for this rare tumor. Treatment by local excision of the tumor alone has been associated with a higher tumor recurrence rate[8] compared to treatment by radical orchiectomy. This may be due to the implantation of malignant cells at the incision site, or incomplete resection due to microscopic invasive tumor growth at the resection margin.[11] The importance of an inguinal or iliac lymph node dissection after an orchiectomy is still being debated. Currently, lymph node dissection is only advocated in cases of suspected spread of disease (i.e. the presence of enlarged lymph nodes on tumor staging CT scans).

More than 60% of these tumors recur within 2 years.[8] However, according to Jones et al,[10] recurrences are common up to 15 years after the initial diagnosis. Plas and colleagues[8] report that recommended treatment includes a clinical examination and abdominal CT scan every 3 months during the first 2 years of follow-up, then yearly CT scans up to 5 years after surgery, and then life-long surveillance.

Conclusion

Our patient presented at a relatively young age with a malignant mesothelioma of the tunica vaginalis and had no obvious history of asbestos exposure. He remains disease-free 7 years after his primary surgery. His young age at presentation and lack of disseminated disease favor a good prognosis. He continues to be followed closely. □

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