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CASE REPORT

Malignant mesothelioma of tunica vaginalis testis: Report of a very rare case with review of the literature

Emanuela Trenti [1], Salvatore Mario Palermo [1], Carolina D'Elia [1], Evi Comploj [1, 2], Alexander Pycha [3], Rodolfo Carella [4], Armin Pycha [1, 5]

[1] General Hospital of Bolzano, Department of Urology, Bolzano, Italy;
[2] Department of Research, College of Health Care Professions Claudiana, Bolzano, Italy;
[3] Kantons Hospital Luzern, Department of Urology, Luzern, Switzerland;
[4] General Hospital of Bolzano, Department of Pathology, Bolzano, Italy;
[5] Chair of Urology, Sigmund Freud University Medical School, Vienna, Austria.

Summary *Introduction: Mesothelioma of the tunica vaginalis testis is a extremely rare tumor and represents 0.3 to 0.5% of all malignant mesotheliomas. Exposure to asbestos often precedes illness. Because of its low incidence and nonspecific clinical presentation, it is mostly diagnosed accidentally during surgery for other reasons and the prognosis is usually poor. We present a case of a patient with a mesothelioma of tunica vaginalis testis, diagnosed secondarily during hydrocele surgery, with long-term survival after radical surgery.*

Materials and methods: a 40 years old patient was admitted to our department for routine surgery of a left hydrocele. During the operation a frozen section analysis was requested because of the unusual nodular thickening of the tunica vaginalis: the examination revealed a diffuse malignant mesothelioma with epithelioid structure and tubular-papillary proliferation. Therefore a left hemi-scrotectomy with left inguinal lymph node dissection was performed.

Results: The definitive histology confirmed the previous report of diffuse malignant mesothelioma with angio-invasion but normal testicle findings and negative lymph nodes. No metastases were found on the CT-scan. For the first 2 years a CT was repeated every 4 months, for other 3 years every 6 months and then yearly. Six years after surgery the patient is classified as no evidence of disease.

Conclusions: malignant mesothelioma of the tunica vaginalis testis is a rare entity, often initially thought to be a hydrocele or an epididymal cyst. An aggressive approach with hemiscrotectomy with or without inguinal and retroperitoneal lymphadenectomy can reduce the risk of recurrence.

KEY WORDS: Mesothelioma; Tunica vaginalis testis; Asbestos exposure.

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INTRODUCTION

Mesothelioma of the tunica vaginalis testis is an extremely rare tumor and the most unusual type, representing 0.3% to 5% of all malignant mesotheliomas. To date only a limited number of cases (about 300) have been reported worldwide in the literature (1, 2). Exposure to asbestos is a well-known risk factor for development of mesothelioma with a long latency between exposure and diagnosis, however, in most cases of mesothelioma of tunica vaginalis testis no asbestos exposure can be documented (2-4). It occurs mostly in middle-aged men but the range of age at presentation can be wide. Because of its low incidence and nonspecific clinical presentation, it is mostly diagnosed accidentally during surgery and the prognosis is usually poor.

We present a case of a patient with a malignant mesothelioma of tunica vaginalis testis, diagnosed secondarily during hydrocele surgery, with long-term survival after radical surgery.

CASE REPORT

A 40 years old patient was admitted to our department for routine left hydrocele surgery. The patient reported progressive scrotal enlargement with discomfort in the left testis and strong groin pain after extended periods of sitting. His past medical history was not significant. No cigarette smoking, trauma or infections were reported. The ultrasonography showed a simple left hydrocele with 350 ml in volume and normal testicular parenchyma. The contralateral testis was normal. The patient underwent resection of the hydrocele; the hydrocele fluid was citrine but the surgeon noted a strange fibrotic thickening of the tunica vaginalis and a frozen section was requested. The patient was discharged one day after the operation, waiting for the definitive histology.

The histologic examination revealed a diffuse malignant mesothelioma with epithelioid structure and tubular-papillary proliferation. The *computed tomography* (CT) showed absence of distant metastases with modest enlargement of the left inguinal lymph nodes up to 22 mm. The patient agreed to a left hemiscrotectomy with left inguinal lymph node dissection, which was thereafter performed. The definitive histology confirmed a diffuse malignant mesothelioma with multiple areas of residual tumors in the tunica vaginalis testis with angioinvasion and stromal infiltration (Figures 1, 2) but normal testicular findings and negative lymph nodes. The immunohistochemical study was positive for calretinin, cytocheratin 5/6, Thrombomodulin, WT1 and D240 while carcinoembryonic antigen and cytocheratin 20 were negative. After consultation with medical and

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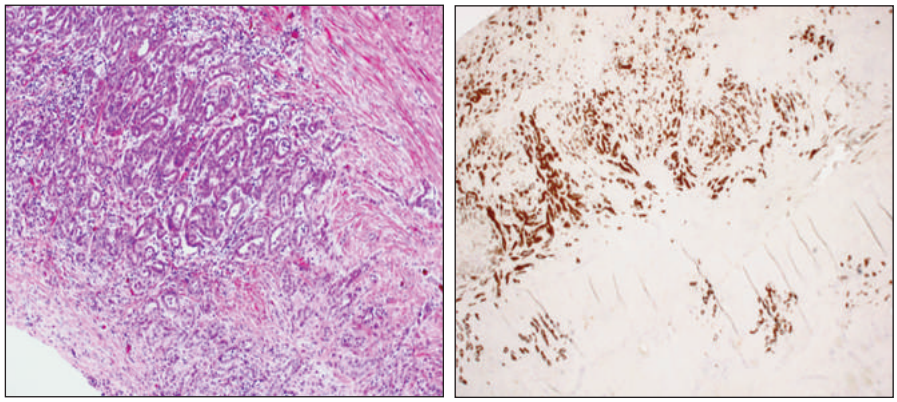

Figure 1.
Neoplastic cells with epithelioid structure and tubular-papillary proliferation with parietal and stromal infiltration (hematoxillin and eosin staining and cytocheratin 5 staining - 4 x magnification).

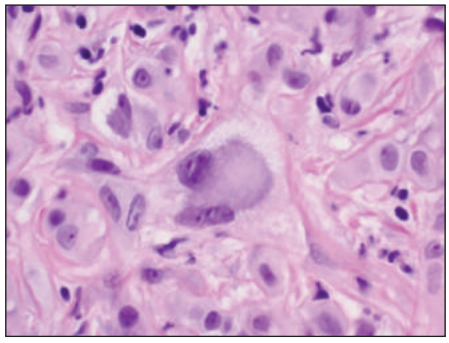

Figure 2.
Mitosis and nuclear polymorphism (hematoxylin and eosin staining - 10 x magnification).

radiation oncologists and in absence of evidence of residual disease, adjuvant therapy was not indicated in our patient. A CT was repeated every 4 months for the first 2 years and every 6 months for the next 3 years, thereafter annually. Six years after surgery the patient shows no signs of recurrent disease. The occupational physician couldn't demonstrate an exposure to asbestos of this patient.

Discussion

Mesothelioma is an extremely rare malignant tumor, which develops from the internal surface of the pleura, pericardium, peritoneum and tunica vaginalis testis. Less then 5% of cases of malignant mesothelioma occur in the tunica vaginalis (5).

The first case was described by *Barbera and Rubino* in 1957 (6).

Exposure to asbestos is a well-known risk factor for development of pleural and peritoneal mesothelioma with a very long latency between exposure and diagnosis, however, exposure is less frequently associated with pericardium and tunica vaginalis testis. Due to its low incidence, it is unknown whether asbestos exposure plays a role in its etiology: less than half of reported mesothelioma of tunica vaginalis testis are associated with asbestos exposure (7). The first case of malignant mesothelioma of the tunica vaginalis testis, associated with asbestos exposure, was reported by *Fliegel* in 1976 (8). In a general review of 223 cases in 2010, *Bisceglia et al.* found an association with asbestos exposure in only 30-40% of the patients (3). Nevertheless in the series of *Spiess et al.* (5) the correlation with asbestos was documented in 80% of the cases and in a recent Italian study, based on the data from the *Lombardy Mesothelioma Registry*, Mensi found an asbestos exposure in 67% of the patients with mesothelioma of the tunica vaginalis testis: here the author underlines the importance to collect the occupational history, the living habits, the residential history and the hobbies of the patients (9).

In our opinion the investigation of the exposure history should be conducted by an experienced occupational physician (2, 3).

Other suspected causes of this kind of mesothelioma are scrotal trauma, long-term hydrocele, herniorraphy and exposure radiotherapy (2-10-11). The age at presentation varies from 7 to 87 years in different reports (2, 4). Because of the lack of characteristic symptoms, these tumors could be confused on clinical assessment with hydrocele or an epididymal cyst and could initially be treated conservatively, delaying the diagnosis.

The patient consults his physician usually for scrotal enlargement, scrotal/inguinal mass or scrotal pain and undergoes surgery with preoperative diagnosis of hydrocele, testicular tumors, inguinal hernia or epididymal cyst. Preoperative testicular ultrasonography could show a nodular thickening of the tunica vaginalis testis and a dense fluid inside but it is mostly negative. Thus, the diagnosis usually occurs secondarily during surgery and the patient needs further surgical treatment: one third of patients, who underwent only hydrocelectomy, experienced local recurrence compared to approximately 11% of patients, who underwent radical orchiectomy (12-13). Inguinal orchiectomy or hemiscrotectomy with inguinal and retroperitoneal lymph node dissection in case of lymph node enlargement and appears to be the preferred treatment for these patients. It is associated with better prognosis and should be proposed when possible. Radiotherapy and chemotherapy have failed to yield significant results and their role is still controversial; however adjuvant radiotherapy could be considered to prevent local disease recurrence while adjuvant chemotherapy with combination of permetrexed and cisplatin, which have had a proven efficacy in pleural mesothelioma, should be considered in cases with unfavorable prognosis (4).

Approximately one third of tumors is locally invasive when diagnosed (1) and more than 50% of patients develop local or distant recurrence with more than 60%

recurrences within the first 2 years (4, 5, 12, 14). The disease specific survival ranges in different studies between 20 and 30 months with 40% of the patients dying from their disease (5, 12, 14) though the last recent series of *Recabal et al.*, with a cohort of 15 patients treated with aggressive surgical management, shows better results: after a median follow-up of 42 months the median overall survival has not been reached (4). Because of the high rate of recurrence a close follow up for the first 2 years is paramount; however a local recurrence may occur up to 15 years after surgery, which is why also a long-life follow up has to be considered (15).

Conclusions

Malignant mesothelioma of the tunica vaginalis testis is a very rare entity, often initially misinterpreted as a hydrocele or an epididymal cyst. Our case shows the importance of a correct diagnosis, even if intraoperatively.
A mesothelioma of tunica vaginalis testis should always be suspected in patients with asbestos exposure and rapid enlargement of hemiscrotum and must always be considered in case of fibrotic or nodular thickening of the tunica vaginalis or in case of hemorrhagic or yellow hydrocele fluid. An aggressive surgical approach with hemiscrotectomy with or without inguinal and retroperitoneal lymphadenectomy can reduce the risk of recurrence and improve the poor prognosis of these patients. A close and life-long follow up is recommended.

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Correspondence

Emanuela Trenti, MD (Corresponding Author)
emanuela.trenti@sabes.it

Salvatore Mario Palermo, MD
salvatore.palermo@sabes.it

Carolina D'Elia, MD
carolina.delia@sabes.it

Evi Comploj, MD
evi.comploj@sabes.it

Rodolfo Carella, MD
rodolfo.carella@sabes.it

Armin Pycha, MD
armin.pycha@sabes.it

Ospedale di Bolzano, via L. Boehler n. 5, 39100, Bolzano, Italy

Alexander Pycha, MD
alexander.pycha@sabes.it

Luzerner Kantonsspital, Spitalstrasse 6000, Luzern 16, Switzerland