

Paratesticular Mesothelioma in Young Age. Case Report

Paratestikulárny mezotelióm v mladom veku. Kazuistika

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Summary

Background: Malignant mesothelioma is a neoplasm arising in serosal membranes in the body cavities. Usual presentation of the tumor is in the pleura, peritoneum and less frequently pericardium. Paratesticular mesothelioma is the rarest known form of malignant mesothelioma with only a limited number of reported cases. **Case:** A case of malignant epithelial mesothelioma of the tunica vaginalis testis was reported with presentation of hydrocele and multinodular intrascrotal masses in a 20-year old male. The patient underwent surgery for post-traumatic long-term hydrocele with perioperative discovery of multiple small exophytic structures. After histological findings of the malignant mesothelioma of tunica vaginalis testis, he underwent left-side orchiectomy, followed by inguinal and pelvic lymph node dissection. Clinical staging did not reveal distant metastases. Regular follow-up visits based on physical examinations and imaging studies are to date negative for recurrence. **Conclusion:** Literature data were reviewed, and possible risk factors for the development of the neoplasm were analysed.

Key words

paratesticular mesothelioma – tunica vaginalis testis – orchiectomy – asbestos

Súhrn

Východiská: Malígný mezotelióm je novotvar vychádzajúci zo serózných blán vystielajúcich telesné dutiny. Zvyčajne sa nádor vyskytuje na pleure, peritoneu a menej často perikarde. Paratestikulárny mezotelióm je najzácnejšou formou malígneho mezoteliómu s obmedzeným množstvom publikovaných prípadov. **Kazuistika:** Autori popisujú prípad malígneho epiteliálneho mezoteliómu tunica vaginalis testis u 20-ročného muža s prítomnosťou hydrokély a mnohopočetných uzlovitých intraskrotálnych útvarov. Dôvodom operácie pacienta bola dlhotrvajúca poúrazová hydrokéla, peri-operačne sa zistil výskyt mnohopočetných drobných exofytických útvarov. Po histologickom diagnostikovaní malígneho mezoteliómu tunica vaginalis testis pacient podstúpil ľavostrannú orchiektómiu s následnou inguinálnou a panvovou lymfadenektómiou. Klinické vyšetrenia neodhalili prítomnosť vzdialených metastáz. Pravidelné kontroly pacienta, pomocou fyzikálneho vyšetrenia a zobrazovacích metód, do súčasnosti nepreukázali rekurenciu ochorenia. **Záver:** Po spracovaní literárnych údajov autori analyzujú rizikové faktory, ktoré mohli viesť k vzniku ochorenia.

Klíčové slová

paratestikulárny mezotelióm – tunica vaginalis testis – orchiektómia – azbest

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Introduction

Malignant mesothelioma is a neoplasm arising in body cavities lined by mesothelium. A type of malignant mesothelioma is based on the section of the mesothelium that it affects. The majority of these tumors are found in the pleura, peritoneum and less frequently pericardium. Paratesticular mesothelioma is the most rare known form of malignant mesothelioma, first described by Barbera and Rubino in 1957, accounting around 0.3–1.4% of all malignant mesotheliomas [1]. To date, less than 250 cases of this type of mesothelioma have been reported worldwide. Asbestos exposure is a major known risk factor in pathogenesis of the tumor. Possible association between radiation therapy and subsequent development of malignant mesothelioma is also suggested. Most of the paratesticular mesotheliomas affect the section of the mesothelium of the tunica vaginalis testis, there are only a few cases of tumors arising in the meso-spermatic cord and epididymis. In the literature review most patients are older than 50 years at the time of their diagnosis. Only about 10% of diagnosed males were younger than 25 years and less than 7% under the age of 20 [1,2]. Patients in general present with a hydrocele or hernia, sometimes accompanied with acute pain syndrome. Palpable masses in scrotum with an insignificant accumulation of serous fluid are also possible. Testes of both sides are equally affected and bilateral or simultaneous tunical involvement has also been observed. A history of asbestos exposure is found in 35–40% of these cases [1,3,4]. Diagnosis is predominantly postoperative, determined by pathologic examination. Clinical differential diagnosis consists of hydrocele, testicular tumors, inflammatory process such as epididymitis, inguinal and scrotal hernia or peritoneal mesothelioma in hernia. There are not characteristic features that can help to differentiate the tumor by clinical examination [5]. Imaging studies can help to identify tunical surface irregularities, soft tissue masses and fluid accumulation. Conventional ultrasound and Doppler ultrasound are most appropriate imaging modalities for primary

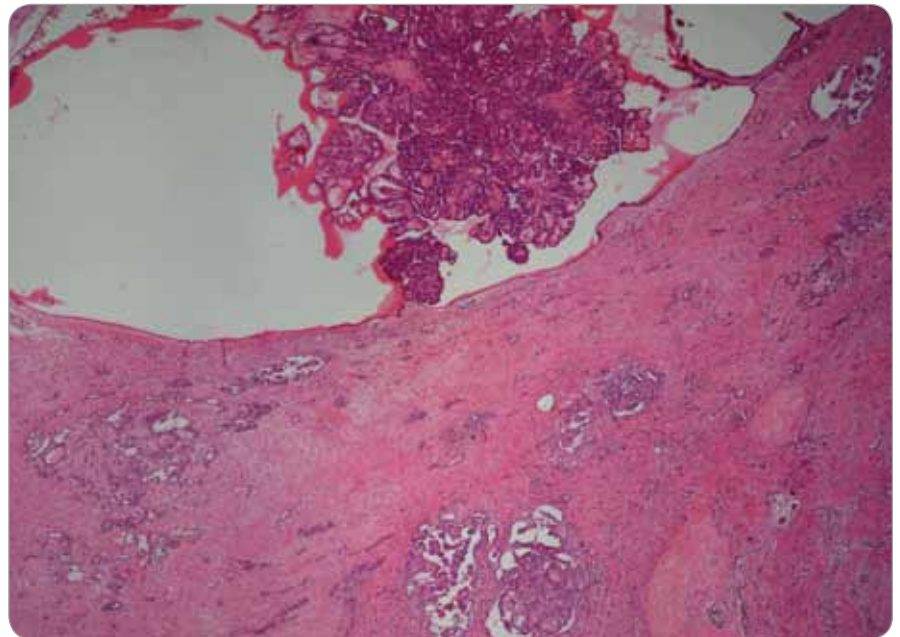


Fig. 1. Histological features of the malignant mesothelioma. Papillary formations are seen in the upper part of the picture, and infiltrative structures are visible in its lower part (H&E; magnification 40×).

tumor, whereas CT and MRI can help to discover lymph node involvement (predominantly paraaortal or pelvic) and distant metastases [1,6]. According to literature data the Tc-99m heat-denatured red blood cell scintigraphy can also reveal the primary tumor [7]. Invasive

diagnostic procedures were not performed frequently in published cases. Aspiration of hydrocele fluid can reveal malignant cells, ultrasound guided FNA of visible masses is more accurate [6,8,9]. There were cases of preoperative diagnosis based on fine needle aspiration

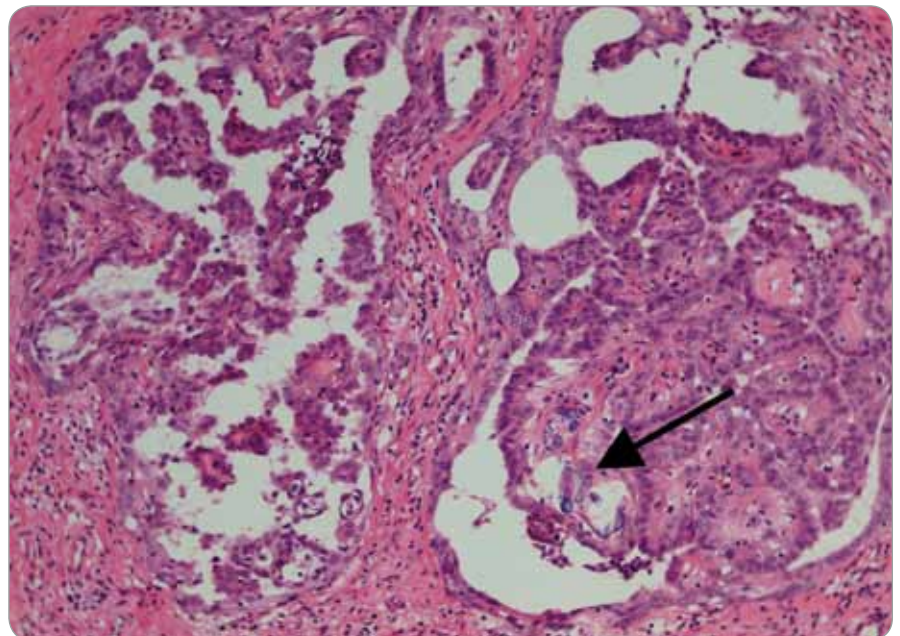


Fig. 2. Detail on the tubular and papillary structures with psammomatous bodies (arrow). (H&E; magnification 200×).

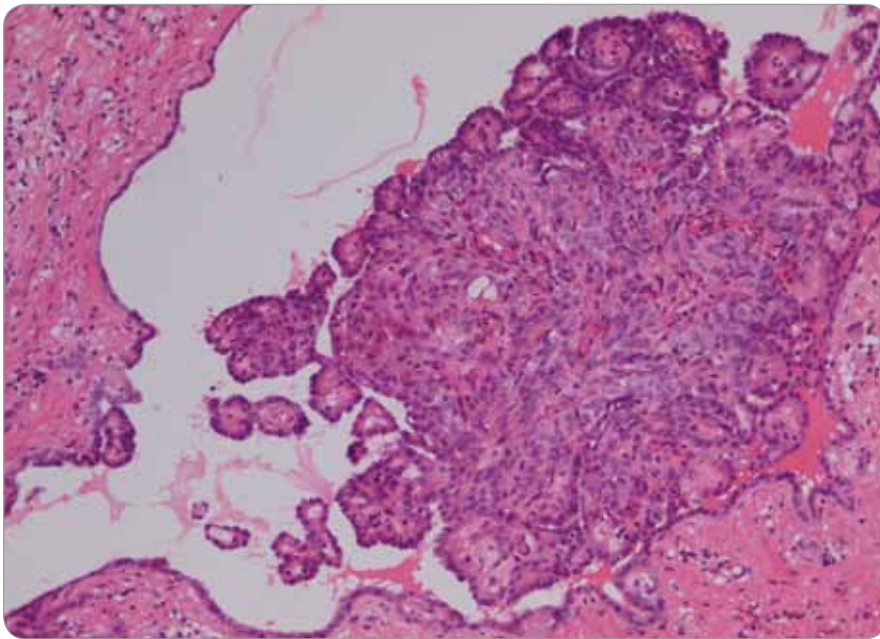


Fig. 3. Detail on the papillary formations. (H&E; magnification 200x).

of the retroperitoneal lymph node metastases [1]. Histo-pathological features of paratesticular mesotheliomas are similar to pleural or peritoneal [10]. In general tumors are epithelial, sarcomatous or mixed. Epithelial types are the most frequent while pure sarcomatous types are very rare. Invasion of subnival connective tissue, testis, epididymis

and other intrascrotal structures is documented in one third of the cases [1,11]. Paratesticular malignant mesotheliomas have capability of local spread as well as lymphatic and hematogenous metastatic potential. Recurrence occurs in 60% of cases within two years, with median survival of 24 months [1]. The overall prognosis is slightly better in com-

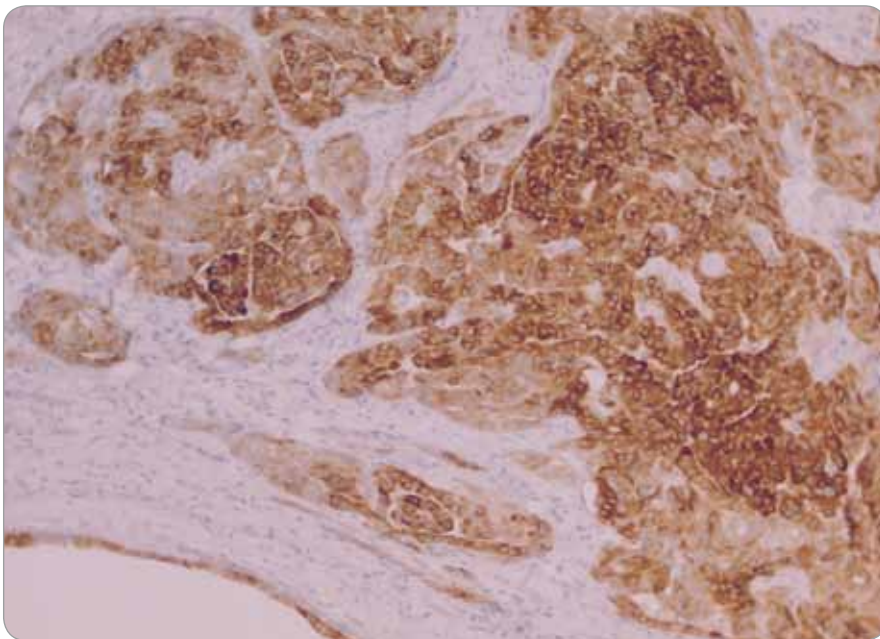


Fig. 4. Immunohistochemical detection of calretinin in the tumorous cells (magnification 400x).

parison with other mesothelioma types, probably because of the early symptom presentation and availability of effective surgical intervention. The aggressiveness of this tumor type requires the first line of surgical treatment – radical inguinal orchiectomy with a lesser local recurrence rate than a more conservative approach [1,12,13]. Systemic treatment such as chemotherapy shows only a little effectiveness. Radiation therapy was also performed as stated in various case studies. Unfortunately reliable guidelines can not be determined due to very limited number of patients.

Case Presentation

20-year old man presented to a regional department of urology with a 9 months history of painless hydrocele of the left testis. Patient had a history of severe motorcycle accident with the injury of the lower extremities and pelvic area three months before the onset of symptoms, history of an infection was denied. Diagnosis of post-traumatic hydrocele was suspected at the time of patient's admission to the hospital. Physical examination revealed left-side hydrocele, his right testis was normal. No lymph nodes were palpable in the inguinal regions. Laboratory findings were completely within reference values, including testicular tumor markers, AFP and β hCG. Marsupialization of hydrocele was indicated. Peri-operatively a rupture of tunica albuginea had happened and revealed numerous small exophytic structures that were sampled by the surgeon and sent for pathological analysis. Histological findings were suggestive of an epithelial type of malignant mesothelioma. Tumor cells were growing in tubular and papillary growth pattern and occasional psammomatous bodies were present (Fig. 1–3). In addition, immunohistochemical profile confirmed the mesothelial origin of the tumor cells because they showed positivity for cytokeratin 7, vimentin, EMA, calretinin (Fig. 4) and were negative for CEA, inhibin, oestrogen and progesterone receptor.

MRI that was performed describes residual fluid collection in post-operative region and minimal right-side hydrocele. Laterally to left testis in soft tissues of the

scrotum multiple small hyperintensive areas were visualised. Displayed inguinal and pelvic lymph nodes were oval shaped, 17–22 mm in diameter, there was no retroperitoneal lymphadenopathy described. Surgical revision was performed three months later including left-side inguinal orchiectomy, inguinal and pelvic lymph node dissection. Histology confirmed the diagnosis of malignant mesothelioma with epithelial characteristics and infiltrative growth in tunica vaginalis. In addition, foci of regressive changes such as giant cells granulomas and old haemorrhage were present. Pelvic lymph nodes were negative for tumor cells, reactive histiocytosis was identified in inguinal lymph nodes, a cluster of tumor cells was found outside the nodes in inguinal fat tissue. The material was sent for second opinion examination that confirmed diagnosis. Post-operative staging CT of the chest, abdomen and pelvis revealed no evidence of metastasis. In paraaortal region marginal multiplication of the lymph nodes was described, individual nodes were under a suspicious size. Laboratory findings remained completely within reference values, including multiple tumor markers. Post-operative radiotherapy and chemotherapy in patient with no evidence of residual disease was not indicated. Regular follow-up every 6 months with physical examination and imaging studies are to date relapse free and patient remains in complete remission after surgical treatment for 41 months.

Discussion

Malignant mesothelioma of the tunica vaginalis is extremely rare primary tumor. The nonspecific symptoms, limited experiences with ultrasound imagining of the mesothelioma features in paratesticular region and wide range of more common diseases in differential diagnosis are probably the reason of minimal pre-operative suspicion of mesothelioma. Diagnosis is almost exclusively accidental. Definitive diagnosis of malignant mesothelioma is possible only on bioptical examination and the differential diagnosis includes reactive mesothelial hyper-

plasia, rete testis adenocarcinoma, ovarian type (serous) carcinoma, metastatic adenocarcinoma and epididymal carcinoma [14]. Malignant mesothelioma may be distinguished from other entities on macroscopic and microscopic features in most cases. Immunohistochemistry plays helping role in challenging cases, for example calretinin is one of the mesothelioma-related markers, which is not expressed in carcinomas of paratesticular localisation. Paratesticular malignant mesothelioma is characteristic of forming ill-defined neoplastic tissue coating or involving the tunica vaginalis in multifocal fashion. On the other hand, rete testis carcinoma and epididymal carcinoma are usually forming solitary mass with destructive infiltration of paratesticular and testicular tissue. Microscopically, architectural features, as complex tubulopapillary proliferation with extensive destruction and confluent infiltration and marked cytologic atypia, are typical for malignant mesothelioma in contrast to reactive mesothelial hyperplasia. Metastatic carcinomas are frequently bilateral and multifocal, and characteristically associated with vascular-lymphatic and interstitial invasion, in patients with a history of primary carcinoma. Most patients with paratesticular mesothelioma are older than 50 years at the time of diagnosis and only less than 7% of diagnosed patients are under age of 20 such as described case [1]. While long-term hydrocele, trauma and hernia are suspected risk factors for developing paratesticular mesothelioma, the only causative risk factor remains asbestos exposure [1,3,4]. There is currently no theory to explain why asbestos exposure might cause a primary tumor to develop in the paratesticular region but we assume that asbestos fibers can deposit in various organs after exposure and result in the development of mesothelioma. After the confirmation of the diagnosis in presented patient, who had a history of both trauma and long-term hydrocele, we also targeted a search for asbestos exposure. We found a link to illegal asbestos dump near patient's habitation, moreover it was located near the

well used for drinking water. This was reported to the legal authorities. First line surgical treatment usually performed as radical inguinal orchiectomy in localized disease plays determinative role in prognosis of patient. Chemotherapy and radiotherapy shows only a little effectiveness and there is understandable lack of evidence-based data for adjuvant treatment. In our case we decided for the regular follow-up based on physical examination and CT imagining with contrast agent. Various markers for mesothelioma such as MesoMerck can be tested. Unfortunately reliable guidelines can not be determined due to very limited number of patients.

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