CASE REPORT

Giant retrovesical epithelioid hemangioendothelioma presenting with acute urinary retention: a case report

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Epithelioid hemangioendothelioma (EH) is a very rare vascular tumor of intermediate biological behavior. It can arise from various sites including bone, liver, spleen, pleura, lung, skin, or soft tissue. We report a case of a 24-year-old man who presented with a history of acute urinary retention and a palpable lump in the lower abdomen. Imaging showed a large pelvic mass displaying heterogeneous texture with solid and cystic components compressing and displacing adjacent structures. The patient underwent exploratory laparotomy with excision of the pelvic mass. Histopathological examination confirmed the diagnosis of EH. Pelvic soft tissue origin of EH is extremely rare. The diagnosis of EH may be suggested by characteristic magnetic resonance imaging (MRI) findings.

Key Words: hemangioendothelioma, acute urinary retention

Introduction

Epithelioid hemangioendothelioma (EH), a rare vascular tumor of intermediate biological behavior, was first described by Weiss and Enzinger in 1982.[1] It usually affects young individuals and involves soft tissue, bone, liver, lung, pleura, breast, brain, or meninges. Soft tissue EHs are usually closely associated with a vessel (often a vein) from which they originate.[1] Imaging or preoperative biopsy does not differentiate between types of vascular tumors including angiosarcoma. The final diagnosis is established only after surgery, as in our case. It is not possible to predict the outcome of this tumor based on histological characteristics, and therefore, it is difficult to make rigid therapeutic recommendations about patient treatment and follow up.

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Case report

A 24-year-old male presented with refractory urinary retention and a lower abdominal lump. Physical examination revealed a large, nontender swelling in the hypogastrium. Rectal examination revealed a nontender, smooth, firm mass bulging into the anterior rectal wall above the prostate. Results from hematological and biochemical laboratory investigations were normal. A computed tomography (CT) scan revealed a 15 cm x 13 cm oval, lobulated mass in the pelvis and pre-sacral region extending inferiorly up to the prostate. It had an intensely enhancing soft tissue component anteroinferiorly and a multiseptate cystic component superiorly. The posterior vesical wall appeared to be infiltrated. A magnetic resonance imaging (MRI) scan of the pelvis showed a 14 cm x 13 cm mass with a heterogeneous texture and solid as well as cystic components. The solid component had an intermediate signal on T1 weighted images and a hyperintense signal on T2 weighted images. The cystic component appeared hypointense on T1 weighted images and hyperintense on T2 weighted images and showed internal septations. Linear intramatrix flow voids were seen within the mass, representing vessels, Figure 1. The urinary bladder was compressed and displaced anterolaterally towards the left. Perivesical fat planes appeared partially obliterated, Figure 2.

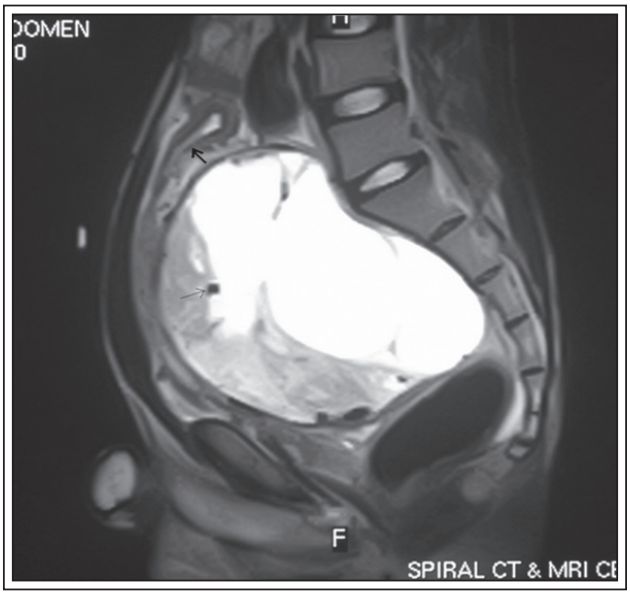

Figure 1. MRI T2–weighted sagittal image showing hyperintense heterogeneous mass (thick arrow) with large intra-tumoral vessels appearing as signal voids (thick arrow) displacing bladder anterosuperiorly with obliteration of flat planes with bladder.

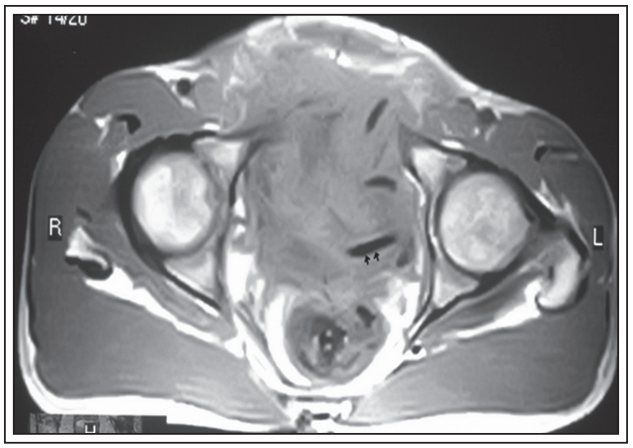

Figure 2. T1–weighted image showing hypointense mass with linear intra-matrix flow voids (arrow indicating flow voids).

Preoperative fine needle aspiration cytology (FNAC) showed the presence of spindle cells, some appearing to be plump fibroblasts. The cells were predominantly clustered with minimal pleomorphism. A diagnosis of soft tissue sarcoma was made, and the patient underwent an exploratory laparotomy using a midline transperitoneal approach. The surgery revealed a hypervascular retrovesical mass with multiple parasitic vessels studded on its surface, infiltrating into the posterior bladder wall and lower 5 cm of the right ureter. The liver and the other intra-abdominal viscera were essentially normal.

We encountered incessant bleeding during mobilization of the tumor. Therefore, a bilateral internal

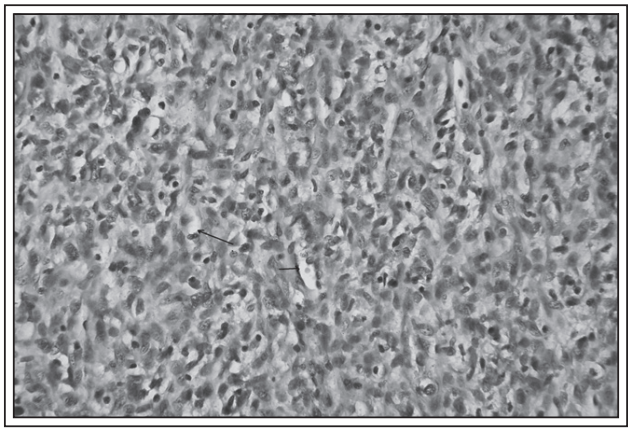

Figure 3. High power photomicrograph showing highly vascularised cellular tumor with moderate degree of nuclear pleomorphism. Many abortive vascular spaces also seen (indicated with arrows) (H&E X450) .

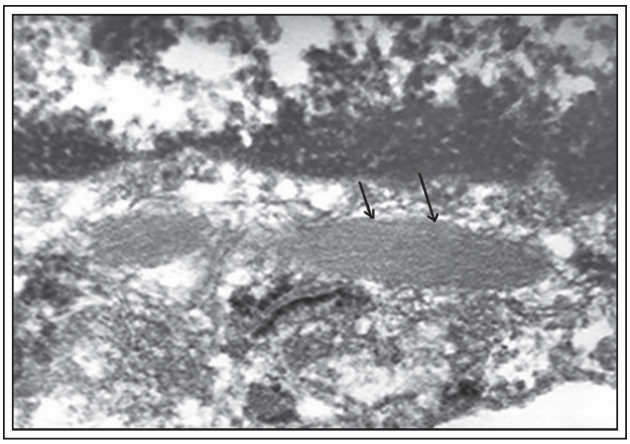

Figure 4. Transmission electron microscopy of a tumor cell showing a cytoplasmic cylindrical structure of a Weibel-Palade body (X28000).

iliac artery ligation was performed. As a result, tumor vascularity decreased significantly. The mass, on mobilization, was found to be free from the rectum, but the posterior bladder wall had to be sacrificed along with the right lower ureter due to tumor infiltration. We performed excision of the mass *in toto*, followed by right ureteric reimplantation and two layer repair of the bladder.

Gross examination of the tumor showed the mass to be solid, lobulated, and unencapsulated. The cut section revealed soft fleshy areas with intervening large hemorrhagic lakes. Histopathological examination revealed a cellular tumor with intervening hypocellular and hypercellular regions. The tumor adhered to the adventitia of a large, muscular blood vessel suggesting its possible site of origin. The tumor cells were round to oval with abundant pale eosinophilic cytoplasm and a moderate degree of nuclear atypia with occasional atypical mitotic activity. The tumor cells lined small vascular spaces that had many abortive vascular channels, Figure 3. Immunohistochemical staining with anti-CD34 showed strong staining by the cytoplasm and membrane, suggesting an endothelial origin of the tumor. Electron microscopy showed characteristic Weibel-Palade bodies, Figure 4, which confirmed the diagnosis of EH. At 6 months, the patient was doing well and was symptom free. The patient was not impotent.

Discussion

Hemangioendothelioma is a distinctive vascular tumor often arising from a vessel. This tumor is intermediate in aggressiveness and can be either benign or malignant.[1] It has several subtypes, including epithelioid, spindle cell, and kaposiform types, as well as malignant endovascular papillary angioendothelioma.[1] EH is characterized by the presence of epithelioid endothelial cells. It occurs in young patients and involves soft tissue more commonly than bone. Soft tissue EH usually occurs in deep tissues of the extremities.[1] One probable cause for these tumors, especially hepatic EH, is exposure to thorium dioxide.[2] Pulmonary EH may be secondary to asbestos exposure.

This tumor often presents with compressive symptoms and pain, due to local tumor expansion. It can also present with bleeding in the target organ.[3] In our case, the patient presented with urinary retention and constipation. Infantile hemangioendotheliomas may present with congestive cardiac failure or thrombocytopenia due to pooling of blood in the tumor.[4]

Given the myriad of possible vascular tumors, the question arises whether a preoperative diagnosis can be made with certainty. It is not possible to differentiate between hemangioendothelioma, hemangiopericytoma, and angiosarcoma radiologically. Radiographs of soft tissue lesions usually reveal only a nonspecific mass. Angiograms reveal dense, well circumscribed areas of staining with early draining veins and shunting. Sonographic, CT, and MRI descriptions of soft tissue hemangioendothelioma, hemangiopericytoma, and angiosarcoma are limited. Sometimes, prominent serpentine vessels can be identified, most often in the periphery of the mass, suggesting these diagnoses.[5]

Percutaneous FNAC has not been very successful in establishing a diagnosis of these lesions, possibly due to bleeding and lack of adequate solid tissue for histological diagnosis. There is a risk of severe bleeding and even death.[6] The role of FNAC in correctly categorizing EH tumors is also questionable.[7] FNAC in our case showed the presence of spindle cells and led us to believe that the tumor could be a sarcoma.

EH tumors are usually treated surgically. There are no definitive therapeutic guidelines for treating this rare tumor. Various adjunctive therapies such as radiation, chemotherapy, and interferon therapy have been proposed, especially for unresectable tumors and giant hepatic EH. Sometimes this avoids the need for extensive hepatic resection and subsequent liver transplantation.[8, 9] Preoperative embolization or sclerotherapy have also been tried in giant hemangioendotheliomas to reduce blood loss or improve resectability.[10]

We performed bilateral internal iliac artery ligation, but the decision was made intraoperatively due to the enormous bleeding that was encountered during

mobilization of the tumor. Radical excision including part of the bladder wall was done, as sarcoma was suspected. However, a review of other retrospective cases suggests this option is often exercised when a preoperative diagnosis is lacking. Nearly 50% of patients who undergo bilateral internal iliac artery ligation are rendered impotent.[11] Our patient did not have postoperative erectile dysfunction.

The diagnosis of EH can be made histologically by the presence of epithelioid cells with intracytoplasmic vacuoles, abortive vascular channels, and a lumina that contains erythrocytes.[12] Immunostaining with Factor VIII or CD-34 antigen can suggest the cell of origin.[13] However, confirmation is made by electron microscopy, which shows the presence of Weibel-Palade bodies that are characteristic of endothelium.[14]

The patient in this case has been followed regularly. There have been case reports where EH has led to local recurrence or even metastases.[4,14,15] Therefore, histological characteristics of the tumor cannot be used as sole guidelines for patient follow up.[16] Strict patient follow up should be maintained to detect local or distant tumor recurrence at the earliest possible time. □

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