

Extraskelatal Osteosarcoma of the Omentum with Aggressive Development

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Rezumat

Osteosarcom extrascheletalmental cu evoluție agresivă

Osteosarcoamele extrascheletale (OSES) au fost descrise pentru prima oară de către Wilson în 1941. Acestea sunt forme neoplazice extrem de rare, reprezentând aproximativ 4% din totalul osteosarcoamelor și 1,2% din totalitatea sarcoamelor de țesuturi moi. Prezentăm cazul unei paciente în vârstă de 66 de ani, internată în clinica noastră din cauza unei hemoragii în spațiul retroperitoneal. S-a efectuat verificarea spațiului retroperitoneal și hemostază. Pacienta a fost reoperată din cauza unei obstrucții intestinale postoperatorii. Cauza acesteia din urma a fost infiltrarea intestinală de la o tumoră omentală de mari dimensiuni, determinată histopatologic ca fiind un OSES. Cazul a fost considerat ca fiind un neoplasm inoperabil, pentru care s-a practicat colostomăpaleativă. În literatură am găsit 3 cazuri de OSES cu originea la nivelul omentului.

Cuvinte cheie: osteosarcom extrascheletal, osteosarcom, sarcom de țesut moale, sarcoame

Abstract

For the first time extraskelatal osteosarcomas (ESOS) were described by Wilson in 1941. They are extremely rare neo-

plasms, presenting 4% of all osteosarcomas and 1,2% of all soft-tissue sarcomas. About 300 cases have been reported in the literature up to date. We present a 66-year-old female patient, admitted in the clinic because of acute bleeding in retroperitoneal space. Revision of the retroperitoneal space and haemostasis were performed. The patient was re-operated because postoperative bowel obstruction. The reason for it was intestinal infiltration from large tumor of the omentum, determined histopathologically as ESOS. The case was considered as an unresectable neoplasm so a colostomy was performed. In the literature we found 3 case reports of ESOS, originated from omentum.

Key words: extraskelatal osteosarcoma, osteosarcoma, soft-tissue sarcoma, sarcomas

Introduction

For the first time extraskelatal osteosarcomas (ESOS) were described by Wilson in 1941. (1) They are extremely rare neoplasms, presenting 4% of all osteosarcomas and 1,2% of all soft-tissue sarcomas. (2,3,4) They can be determined as rare mesenchymal tumors characterized by the production of neoplastic osseous tissues without connection to bone or periosteum. (5) They occurs commonly in lower extremities, followed in frequency by upper extremities and retroperitoneal space. Tumor localization in the visceral organs is extremely rare. (6) Since their first description, only about 300 cases have been reported in the literature up to date. (7) ESOS are very aggressive and usually at the time of presentation they are unresectable, with lung metastases.

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Case report

A 66-year-old female patient was admitted in Neurological Department of University Hospital "Queen Joanna" due to vertigo, pain, weakness and sensory hyposensitivity in the right lower extremity. Complaints had appeared a day before the hospitalization. Abdominal pain, hypotension (90/60) and tachycardia (110 beats per minute) appeared after admission of the patient. The pulse was well filled and rhythmic. The laboratory tests revealed Haemoglobin 83 (g/l), Hct 0,34 (l/l), erythrocytes 3,8 ($\times 10^{12}/L$), leucocytes 16,9 ($\times 10^9/L$), platelets 793 ($\times 10^9/L$), albumin 31 (g/L), total protein 61 (g/L), direct bilirubin 25,6 ($\mu\text{mol/L}$), total bilirubin 40,4 ($\mu\text{mol/L}$), AST 83 (IU/L), ALT 27 (IU/L), GGT 102 (IU/L), AF 891 (IU/L). The investigation of the haemostatic status of the patient showed lack of coagulation. The patient was treated with per oral anti-coagulant during the last 2 years after an operation with placing prosthetic aortic and mitral valves. The patient had heart failure III functional class.

The performed abdominal ultrasound was without pathological findings. The CT revealed a retroperitoneal formation in the right side, probably haematoma, scarce effusion in the abdomen and pelvis, osteophytes, osteochondrosis and scoliosis in the lumbar segments of the spine. The patient was admitted in the surgical department and an urgent operation was carried out.

The Intraoperative exploration revealed haemorrhagic effusion about 200 ml. The whole retroperitoneal space in the right side and intestinal mesenterium were imbued with blood, with presence of multiple haematomas. There were suffusions into anterior gastric wall, transversal colon and the right colonic flexure and petechiae on the surface of the rest intestinal loops. The right adnexa were imbued with blood. The omentum had zones of fibrous degeneration and denser structure and was with multiple haemorrhagic petechial changes. The peritoneum was dissected along the right abdominal canal and the retroperitoneal space was opened - a haematoma was found in its right side. The haematoma was spread out to pelvis and imbued the mesenterial radix and the mesenterium of the sigmoid colon. The multiple coagulum were removed but diffuse contact bleeding from each surface was detected. There were no lesions of the aorta, inferior vena cava, iliac and mesenteric vessels. Surgical haemostasis was performed with partial control of the bleeding. The haemostatic sponges and tamponade with gauze tapes were placed. After placement of signal drainages in the abdominal cavity and retroperitoneal space, the laparostomy was performed for second-look operation and lavage. Intensive substitution treatment with blood and blood products and systemic haemostatic therapy were carried out. During the second operation (a day after the first) a source of bleeding was not found, the tamponade was removed and haemostatic sponges were placed again in the retroperitoneal space. It was found that in the omentum there was large suffusion without active bleeding, but with single grey fibrous areas, considered to be result from haemorrhagic infarction. The abdominal cavity was closed. The condition of the patient was gradually stabilized and she was extubated. The laboratory test revealed stable levels of Haemoglobin 103 (g/l),

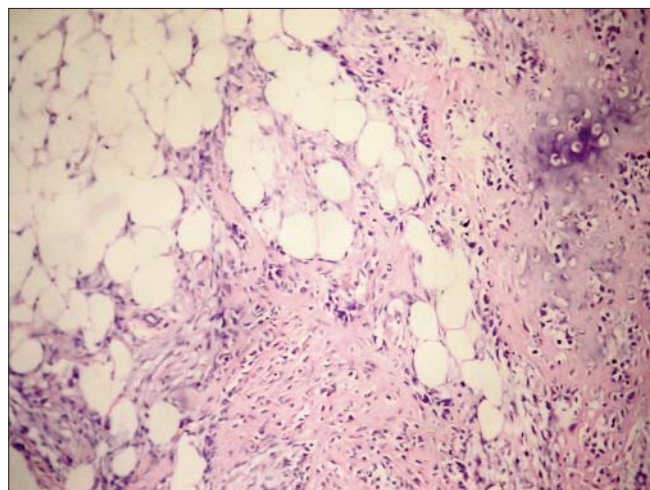


Figure 1. Extraskelatal osteosarcomas

Hct 0,34 (l/l), erythrocytes 3,9 ($\times 10^{12}/L$), but still increased levels of alkaline phosphatase -986 (IU/L). Rehabilitation was started on the fourth day after the operation. Until the 6-th day after the operation the flatulence was not restored, the abdomen was moderately painful and bloated. Because of the intensifying ileus the patient was re-operated on the 7-th post-operative day. The intraoperative exploration revealed tumor transformation of the whole omentum with hard, bone consistency and infiltration of all intestinal loops which cannot be surgically dissected.

The right colonic flexure was mobilized and loop colostomy was performed. Excision biopsy from the omentum was carried out. Histopathological data proved that the tumor was ESOS. (Fig. 1)

The case was discussed by oncologic commission who recommend chemo- and radiotherapy but the patient refused the treatment. The patient was discharged on 9-th post-operative day. She died 3 months later.

Discussion

ESOS can be divided to osteoblastic, fibroblastic, chondroblastic, malignant fibrous histiocytoma-like, telangiectatic and well-differentiated. (8) Primary tumors originated from omentum are rarity. We found only 3 reports of ESOS of the omentum. (9)

Osteosarcomas primarily affect young patients, occurring most frequently before age of 30, with second peak in the fifth and sixth decade. This is more typical of ESOS. There is a slight female predominance. (10)

Osteosarcomas are the most common primary bone tumors occurring usually in the metaphysis of long bones in young patients. (11) ESOS may occur in every part of the body. They most frequently affect lower extremities (42,3%-52%), followed by upper extremities (11,5%), retroperitoneal space (11,5%) and buttocks (7,7%). (4) ESOS have been reported to develop in unusual sites - larynx, kidneys, esophagus, small bowel, liver, heart, urine bladder, salivary gland, breast, etc. (4)

ESOS have been reported to be associated most frequently with trauma, local radiotherapy or malignant fibrous tissue

disease. (12) There are 2 theories about their genesis (13):

1. Tissue residue theory according to which there are mesoblastic residues from the period of embryonic development and bone tissue and osteosarcomas subsequently arise from them;
2. According to the second theory, it has been suggested that muscle interstitial fibroblasts are exposed to external or internal stimulation (trauma, inflammation), undergo metaplasia of the osteoblasts or chondrocytes, which evolves into osteosarcoma.

ESOS contain malignant and primitive spindle cells with varying amounts of osteoid, osseous or cartilaginous tissue. (12) Histopathologically ESOS have 2 microscopic components – sarcomatous cells and extracellular matrix, consisting of osteoid or immature bone tissue. (4)

Osteosarcomas and ESOS are extremely various regarding to immunophenotype by reacting to factor-XIII related antigen, S100, desmin, alpha-smooth muscle actin, cytokeratin and epithelial membrane antigen. (14,15) It has been considered if a tumor produces osteoid or bone nevertheless its localization, it may be classified as osteosarcoma. (16)

Extraosseous metastases most often occur in the lungs (~80%) and kidneys. There are rare cases of metastasis in the heart. (12,10)

Diagnosis of primary ESOS is based on the suggested by Allan et al. criteria (1971) (17):

1. the presence of a uniform morphological pattern of sarcomatous tissue that excludes the possibility of malignant mesenchymoma;
2. the production of malignant osteoid or bone by the sarcomatous tissue;
3. lack of attachment to bone system.

The bone scintigraphy has a main role in initial evaluation of the patients with primary bone tumors or in detection of metastases. (10) X-rays show a fairly clear soft tissue mass with multiple calcifications in the lesion, which is not attached to the surrounding bone structures. (12)

ESOS can be mistaken for myositis ossificans, which is a benign ossifying process occurring in young ages as a well defined masses within the skeletal muscle without cytologic atypia in them. (18)

Only radical resection of the tumor (R0) has curative potential and the wide excision in tumor-free margins is related to better prognosis. (12) Radio- and chemotherapy have not a significant success affecting the final survival. (7,19) Combination of adjuvant and neoadjuvant chemotherapy is without demonstrable increase in disease free survival or overall survival. (19) Brachytherapy has been tried only in the setting of clinical trials. (20) Polychemotherapy based on Adriamycin (adriamycin- ifosfamide) is recommended with priority given to monotherapy. The overall prognosis remains poor, with a 5-year survival rate about 25%-37%. (7,20,21,22,23) Approximately 50% of tumors recur and about 60% of surgically treated patients develop lung metastases. (23) Tumor size, patient's age and localization of the tumor process are important prognostic factors.

Conflict of interest

The authors declare that they have no conflict of interest.

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