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Adamantinoma of the pelvic bone, a difficult diagnosis with fatal outcome

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Abstract Adamantinoma is a primary low grade malignant bone tumor that is predominantly located in the mid-portion of the tibia. The tumor is of interest for two reasons: first, there still exists considerable dispute as to the origin of the lesion and recent reports reveal that the condition is more malignant than had previously been supposed. Although cases of adamantinoma located to the axial skeleton have been reported, this is the first case of adamantinoma located to pelvic bone in Iran. Here we present the clinical, radiological & histopathological features of a 19 year-old male with painful lesion located to the right pelvic bone which was morphologically and immunohistochemically diagnosed as adamantinoma. In general, metastasis is seen in 15%–20% of patients. The spread can occur to regional nodes, lung and infrequently to skeleton, liver and brain [1]. Several weeks after surgery, our patient's condition gradually worsened. A CT-scan of abdomen revealed widespread liver metastasis and the patient died due to acute liver failure. This case demonstrates that the mortality rate from adamantinoma is not always low.

Key words adamantinoma; pelvic bone; diagnosis

Adamantinoma is a primary, low grade, malignant bone tumor of unknown histogenesis. It is a rare neoplasm comprise only 0.1%–0.5% of all primary bone tumor [2]. The first reported example is attributed to Marier in 1900 [3]. In 1913 Fischer [4] named the lesion "primary adamantinoma of the tibia" because of its striking histologic resemblance to the jaw adamantinoma. In general, adamantinoma is a slow growing tumor and tends to be locally aggressive and rarely metastasizes [5]. The rate of metastasis is approximately 15%–20% and usually occurs in the first two years following diagnosis [6, 7]. The most common sites of metastasis are bone, lung and regional lymph nodes [6]. To our knowledge, until now, adamantinoma has been reported only in few locations in the appendicular skeleton. Here we present a patient with an adamantinoma of the pelvic bone who subsequently developed widespread liver metastasis and died due to acute liver failure.

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Case presentation

A 19 year-old Iranian male complained of pain in the right pelvic area and limping from one year ago. The pain persisted and did not decline over the following several months. There was no history of previous trauma. One month ago he consulted an orthopaedist in the community hospital. According to his medical records, active and passive range of motion of right hip joint were reduced, but neurovascular examination were normal. X-ray and CT scan showed slight sclerosis, cortical thickening, cortical destruction and periosteal reaction (vertical spicules) of right iliac bone with soft tissue thickening of gluteus and iliacus muscles (Fig. 1 and 2). Except for alkaline phosphatase which was 700 U/L (normal value = 80–306 U/L), and all of the laboratory data were unremarkable. Needle biopsy from right anterior iliac bone was performed and a definite pathologic diagnosis could not be made from that specimen, but metastatic adenocarcinoma was considered possibility. Extensive metastasis work up including chest and abdominal CT-scans, Tc-sintigraphy, endoscopy, colonoscopy and tumor markers evaluation were negative. Then the patient referred to our center

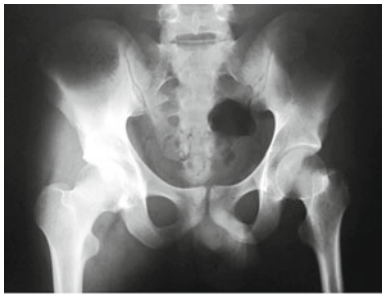

Fig. 1 X-ray showed slight sclerosis, cortical thickening

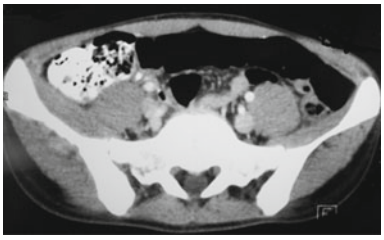

Fig. 2 CT scans showed cortical destruction and periosteal reaction (vertical spicules) of right iliac bone with soft tissue thickening of gluteus and iliacus muscles

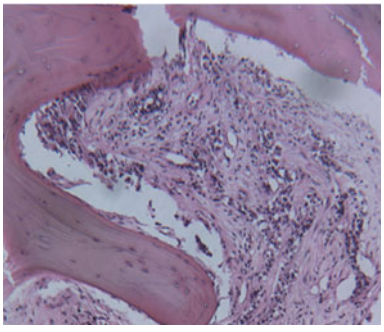

Fig. 3 Microscopic examination showed a biphasic tumor, composed of epithelial and osteofibrous components (× 10)

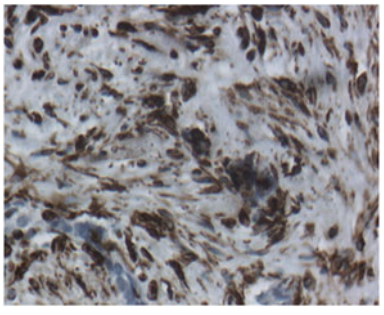

Fig. 4 Epithelial and spindle cells were positive in vimentin staining (× 40)

and underwent open surgery. At surgery, the tumor partly was removed for histologic examination, but a through enucleation was postponded to the next step due to lack of necessary facilities. The resected tissue, which was of hard consistency, paraffin blocks and glass slides related to previous surgery were sent to our department for examination and consultation. Microscopic examination showed a biphasic tumor, composed of epithelial and osteofibrous components. Epithelial cells were medium to large in size with finely dispersed chromatin and an overall bland appearance. Mitotic figures were infrequent. These cells had arranged in nests and tubular or glangular appearance. Islands of epithelial cells were surrounded by spindle cells. The spindle cells had interlacing fascicles and there was no matrix production (Fig. 3). Our diagnosis was classic adamantinoma. IHC studies revealed that the epithelial cells expressed keratins and staining for vimentin was strongly positive in epithelial nests and fibroblast-like stroma (Fig. 4). Tumor cells were negative for S100, CEA and CD99. The diagnosis of adamantinoma was confirmed.

Discussion

This case is of interest because of the lesion located to the pelvic bone and early liver metastasis. Adamantinoma of the long bones is an uncommon, primary low growing malignant bone tumor. Adamantinoma presents a wide variety of histologic type with malignant biologic behavior. Adamantinoma mostly occurs in the second to fifth decade. The mean patient age is 25–35 years with a range from 2 years to 86 years. It is slightly more common in men than women, with a ratio of 5:4 [8]. Our patient was a 19 year-old Iranian male. The tumor has striking predilection for the long bone (97% of cases) and specifically, the tibia (80%–85% of cases). Other bones that are involved in order of decreasing frequency including humorous [9], ulna [10], femur [11], fibula [12], radius [13], innominate bone [14, 15], rib [16], spine [17] and rarely small bones of the hand and foot [18]. The present case is unique in that the patient had a novel site of appearance of adamantinoma. A history of significant trauma has been noted in about 60% of 200 cases reviewed by Moon and Mori [8]. The presented patient had no history of trauma. The initial symptom are often indolent and non specific. Our case complained of pain in the right pelvic area for one year ago. As far as radiologic features are concerned, the most common appearance is that of multiple sharply circumscribed lucent zones of various size, with sclerotic bone interspersed between the zones and extend above and below the lucent zone. Sometimes the radiographic accepts of adamantinoma are similar to the those of osteofibrous dysplasia of tibia [19]. MRI is useful in differentiating between adamantinoma from osteofibrous dysplasia. On plain radiograph, both of them may show polycyclic osteolysis refereed to as soap-bubble appearance. MRI can give information not only on the extend of the tu-

mor, but also about its origin [20]. In the presented case CT scan showed slight sclerosis, cortical thickening, cortical destruction and periosteal reaction (vertical spicules) of right iliac bone with soft tissue thickening of gluteus and iliacus muscles. The main differential diagnosis of adamantinoma is epithelial metastatic neoplasm. However the bland cytological growth along with the soap-bubble like appearance of adamantinoma would be quite unusual for a metastatic epithelial neoplasm. The presented patient underwent extensive metastases work-up which were negative. Adamantinoma are usually treated by excision with wide surgical resection of the tumor and insertion of a segment of intercalary bone allograft or osteoarticular segment [21–23]. Amputation is the treatment of choice in patients presenting with local recurrence, metastases or failure of reconstruction [23]. The tumor appears relatively insensitive to radiotherapy [6] and although the optimal treatment is still controversial, a need for early aggressive surgical treatment has been suggested by some authors [24]. Our patient died before radical surgery. Some studies reported the long term outcome of adamantinoma [25–28]. Male sex, pain, symptoms of < 5 years' duration and initial treatment by biopsy, curettage or resection are risk factors for recurrent or metastatic disease [26]. The rate of distant metastasis in long bone adamantinoma is about 15%–20% [8, 26]. This percentage may be higher because the indolent course of the disease. In 1986 Moon and Mori found 14 metastases in 109 cases [8]. Distant metastases were mainly found in patients with a history of local recurrence, mostly due to inadequate initial treatment. Lungs are most frequently affected, less frequently metastases are found in regional lymph nodes, liver and bones. Szendroi *et al* [28] published a long term follow-up study of 11 cases of adamantinoma of long bones. The authors reported recurrences in 4 patients 20 and 16 years after initial treatment. One patient died of pulmonary metastasis, 9 years after diagnosis. Although adamantinoma is a low grade, slow growing malignant bone tumor, a lifelong follow-up of the patient is necessary due to the possibility of recurrences or metastasis even decades after the primary tumor. Mortality rates of 13% [26] to 18% [8] have been reported. The mean duration of survival with metastatic adamantinoma of long bones is estimated to be approximately 13 years [29]. To our knowledge liver metastasis in adamantinoma is rare and has favorable course [30] but in contrary the presented case is unique in that the patient had not only a novel site of appearance but also had early widespread and fatal liver metastasis. We find that the mortality rate from adamantinoma is not low and early aggressive treatment and long term follow up are mandatory. Recently, the least common variant of adamantinoma has been described as Ewing's sarcoma like adamantinoma or adamantinoma like Ewing's sarcoma. The cells exhibits features of both epithelial cells and neuroendocrine cells. IHC have shown the tumor cells to contain both epithelial and neural antigens [31, 32]. Bridge *et al* [33] documented translucation (11, 21) in the nuclei of cytokeratin-immunoreactive cells. Identification of t (11, 21) by cytogenetic and/or molecular genetic techniques is specific in Ewing's sarcoma, however is not universally available. So IHC techniques are more popular. In about our case the tumor cells were negative for CD 99 and S100 antigens and the cells did not have "round cell" morphology. The histogenesis of this variant is not clear, however Ewing's sarcoma is considered to be derived from primitive pleuripotential stem cells that may differentiate into cells with mesenchymal, epithelial and neural features. In conclusion the diagnosis of adamantinoma requires knowledge of compatible clinical and radiologic studies as well as understanding of the variable histologic patterns that one may encounter. In addition we find that the mortality rate from adamantinoma is not always low and early aggressive treatment and long term follow up are mandatory.

Conclusion

Adamantinoma of the long bones is an uncommon, primary low growing malignant bone tumor. Although cases of adamantinoma located to the axial skeleton have been reported, but it is rare. The diagnosis of adamantinoma requires knowledge of compatible clinical and radiologic studies as well as understanding of the variable histologic patterns that one may encounter. The mortality rate from adamantinoma is not always low and early aggressive treatment and long term follow up are mandatory.

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