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CASE REPORT

CLEAR CELL SARCOMA OF METATARSUS

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СВЕТЛОКЛЕТОЧНАЯ САРКОМА МЕТАТАРЗАЛЬНЫХ КОСТЕЙ

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ABSTRACT

We report a case of clear cell sarcoma in the third metatarsus of the right foot. This type of tumor is very rare and scantily reported in literature. A 42-year-old Caucasian male presented with a nodular ulcerated mass on the dorsal side of the left foot. X-rays demonstrated a nodular solid lesion which dislodged the third metatarsus. A biopsy revealed a neoplastic proliferation with a sarcoma clear cell profile; because of the aggressive nature of this type of neoplasm, we performed a trans-tibial amputation according to Bugess to achieve a better functionality for the patient. The present study underlines clinical, morphological, as well as imaging and therapeutic aspects of a rare neoplasm such as clear cell sarcoma. The location site is also quite unusual - the metatarsus of the foot. The histological and immunohistochemical data were suggestive of the diagnosis of clear cell sarcoma of metatarsus. After MRI and a bone scan, the surgical treatment suggested the extension over the forefoot and the ankle and therefore a trans-tibial amputation was made.

Key words: *metatarsus, clear cell sarcoma, immunohistochemistry, orthopaedic management, amputation*

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РЕЗЮМЕ

Оповещаем о случае светлоклеточной саркомы в третьей метатарзальной кости правой стопы. Данный тип опухоли является исключительно редким и не нашёл должного освещения в литературе.

42-летний белый мужчина поступил на лечение с изъязвлённой узловатой массой на дорзальной стороне левой ноги. Рентгеновское изображение показывает наличие нодулярного твёрдого поражения, вытеснившего третью метатарзальную кость. Биопсия раскрывает неопластическую пролиферацию с характеристиками светлоклеточной саркомы; ввиду агрессивного характера данного типа неоплазмы мы провели транстибиальную ампутацию по методу Bugess для улучшения функционального состояния пациента.

В данном исследовании мы останавливаемся на многообразных клинических, морфологических, образных и клинических аспектах редких типов опухолей из рода светлоклеточной саркомы. Локализация опухоли также является необычайной – метатарзальные кости стопы.

Гистологическое и иммуногистохимическое исследования показали, что речь идёт о светло-клеточной саркоме метатарзальных костей. После проведения магнитно-резонансной томографии и сканирования костей было установлено, что опухоль расширяется над передней частью стопы и лодыжки, вследствие чего была проведена транстибиальная ампутация.

Ключевые слова: *метатарзальные кости, светлоклеточная саркома, иммуногистохимия, ортопедическое лечение, ампутация*

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Metatarsal tumors are rare affecting males more frequently.[1,2] In this site, clear cell sarcoma (CCS) known as "malignant melanoma of soft tissue"[3,4], occurs rarely in tendons, aponeuroses and bones of adults.[5,6] *A 42-year-old Caucasian male was admitted*

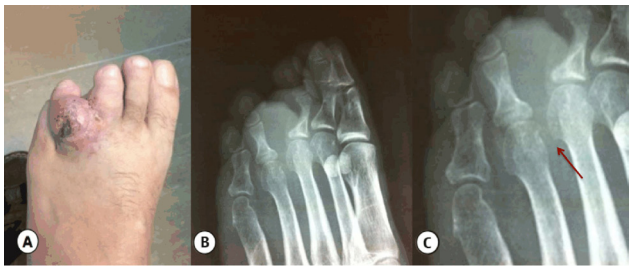

Figure 1. Macroscopic clinical presentation of the neoplasm (A); X-rays showed a dislocation of the third metatarsal bone with osteolytic rim (B); Close-up of the lesion (C).

to the orthopedic outpatient clinic with a 5-month history of pain in the right foot; he related this to a traumatic contusion sustained six months before which was treated medicamentally. At physical examination, a painful, nodular, ulcerated mass was revealed on the dorsal side of the left foot between the third and fourth metatarsus (Fig. 1A). X-rays demonstrated an expansive nodular lesion, which dislodged the third metatarsus showing a thin rim of osteolysis (Figs 1B, 1C).

Incisional biopsy of the lesion was performed and the sample was sent to the pathology laboratory. Grossly, the specimen was a 4.3x3x2.5-cm firm oval mass, gray-whitish in color, difficult to cut; its cut surface was homogeneous and white, showing a partial pseudo-capsule and a small portion of adherent bone. Then, the surgical sample was fixed in 10% neutral buffered formalin for 36-48 hours at room temperature; successively, after a demineralization with 1% in formic acid for 12 hrs, it was embedded in paraffin at 55° C and cut into 4-micron-thick sections mounted on poly-L-lysine coated glass slides. Haematoxylin-eosin routine histological stain was initially performed; on parallel serial sections immunohistochemical procedures were carried out, as previously reported elsewhere.[7] Histologically, the tumor showed neoplastic proliferation represented by sheets of spindle cells, which dissociated the dense connective collagen and was adjacent to bone tissue (Fig. 2); the great majority of neoplastic population was constituted by elements with clear cytoplasm, round or oval nucleus and prominent nucleolus (Figs 3A, 3B).

The immunohistochemical investigations docu-

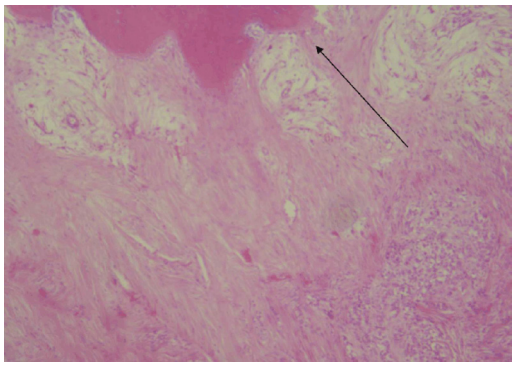

Figure 2. Nests of neoplastic elements infiltrating the adjacent connective tissue and bone (see arrow, haematoxylin-eosin, x100).

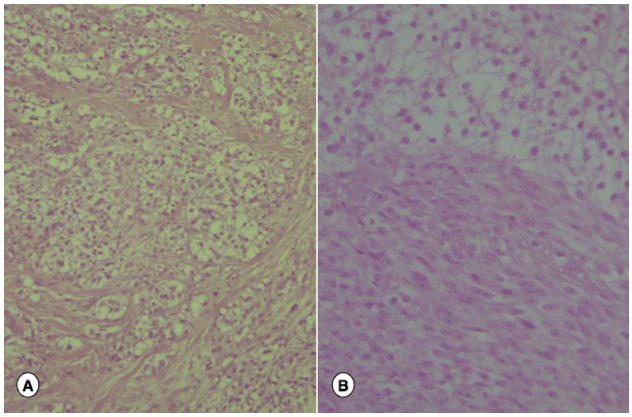

Figure 3. Histological appearance of clear cell sarcoma (A, haematoxylin-eosin, x160); neoplastic clear cells were intermingled with oval, spindle, nucleolated elements (B, haematoxylin-eosin, x220).

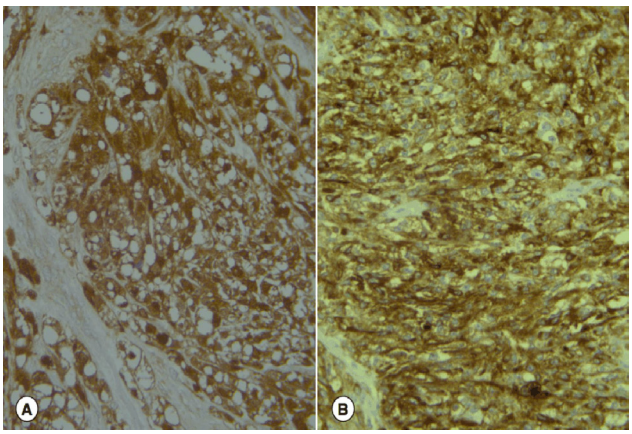

Figure 4. Intense cytoplasmic immunostaining for vimentin was seen in neoplastic clear cells (A) (immunoperoxidase, x200); an evident perinuclear granular immunoreactivity was observed for melan-A (B) (immunoperoxidase, x200).

mented an intense, diffuse or granular, cytoplasmatic positivity for vimentin (Fig. 4A), S100 and melan A (Fig. 4B) in neoplastic elements, while a not

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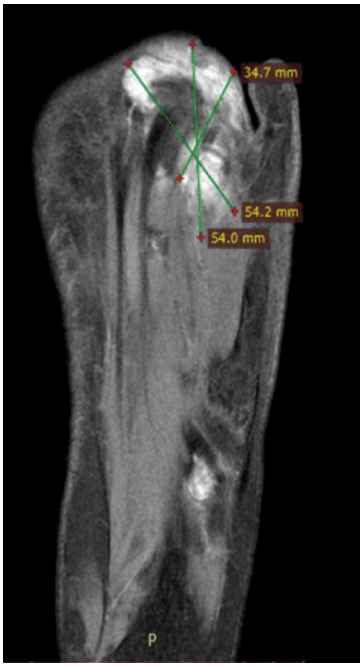

Figure 5. MRI revealed an evident tumor mass.

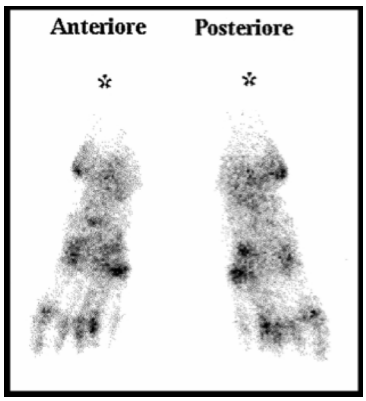

Figure 6. Left foot bone scan with captation by forefoot and rearfoot.

homogeneous immunoreactivity for HMB45 (Human Melanoma Black) and NSE (neurone-specific enolase) was recorded. SMA (Smooth Muscle Actin, desmin, CD117, and CD99 immunostainings were constantly negative as well as broad spectrum cytokeratin AE1/AE3. The histological examination, in association with the immunohistochemical data, lead to the diagnosis of clear cell sarcoma. Tooms technique was performed, cutting at 14 cm from the medial articular edge of the tibiae.

The surgical specimen was initially fixed in 10% neutral buffered formalin for 24 hrs and then subjected to anatomopathological section and the fragments obtained were then demineralized with 1% formic acid for 72-96 hrs; in particular, samples taken from cuboidal and malleolar bones were examined. The histopathological examination confirmed the diagnosis of clear cell sarcoma. In the metatarsus, the most numerous neoplasms are of cartilaginous origin and generally benign, such as enchondroma.[8] Malignant tumours are much rare than the benign ones, representing only 20% of all neoplastic lesions[1,2]; among them, chondrosarcoma and Ewing's sarcoma are considered the most common[2]. CCS, first described by Enzinger (1965), has been considered to have its origin from neural crest cells and a melanocytic differentiation.[3,4] Some reviews concerning CCS occurrence in tendons and aponeuroses has been performed[3,4], while only isolated case reports have shown a primary localization of CCS in bone of young adults[5,6], in these latter cases, at conventional radiographs, CCS showed a lytic lesion of rib, humerus as well as radius, while the metatarsal occurrence was absolutely rare[5,6]. Therefore, because of the unusual localisation of CCS in metatarsus we considered the present case worth reporting. Histological findings of the lesion were greatly suggestive of CCS diagnosis, although only the evident immunoreactivity for vimentin, S100 and melan A as well as the non-uniform immunostaining for HMB45 and NSE strongly confirmed its histogenetic melanocytic origin from the neural crest, as previously suggested elsewhere.[3,4] In the surgical management of this aggressive neoplasm, two choices were possible such as the disarticulation of the ankle or trans-tibial amputation. According to the principle of oncological radicality, mainly considering the presence of cuboidal and malleolar lesions, suspected of being "skip metastases" at MRI and confirmed at bone scan (Figs 5, 6) the surgical treatment suggested the extension over the forefoot and the ankle; it means that the resection edges have to guarantee at least a free margin of 2 cm. Moreover, from the functional and prosthetic point of view, the trans-tibial amputation is more convenient than the disarticulation of the ankle or the amputation of distal leg because it allows larger and more efficient prosthetic options and walking recovery. Finally, the tissues of distal leg are relatively low vascularized and the thickness of soft parts is thin; therefore, even well treated stumps, can lately ulcerate due to the prosthesis and the aging physiological amputation.

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