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CASE REPORT

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Chondrosarcoma secondary to hereditary multiple exostosis treated by extended internal hemiplevectomy

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The authors report on the case of a 28-year-old patient with extensive chondrosarcoma of the left ischium and pubis involving hip joint, skin, and soft tissue of the gluteal region, secondary to hereditary multiple exostosis submitted to an extended internal Enneking type II and III hemipelvectomy. No prosthesis or arthrodesis was used. A few years ago, patients with extensive tumors like this one were treated with interilioabdominal amputation, resulting in a loss of quality of life. Two years after the limb-preserving surgery, this patient was disease free, with good functional results, including bipedal ambulation with support.

UNITERMS: Chondrosarcoma. Osteochondroma. Surgery. Prosthesis. Iliac bone. Osteochondromatosis.

INTRODUCTION

Hereditary multiple exostosis (HME), described by Boyer in 1814,[1] is a dominant autosomal inherited disorder characterized by the presence of multiple osteochondroma. Transformation of HME ranges from 10-38 percent, and in solitary osteochondroma from 1-16 percent.[2] Surgery is the treatment of choice for osteochondroma as well as for chondrosarcoma. This report presents a case of locally advanced chondrosarcoma, secondary to HME. Although a candidate for an interilioabdominal amputation, the patient's limb was preserved with an internal extended hemipelvectomy, without utilization of prosthesis or arthrodesis for the reconstruction. The patient presented good oncological, esthetic and functional results two years postsurgery.

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CASE REPORT

A 28-year-old white male was first seen on April 10, 1994, reporting a tumor in the left gluteal region for the prior five years. He had been submitted to a previous resection elsewhere, and had suffered a local recurrence.

Deambulation was hindered by restriction of hip joint movements. Upon physical examination, the patient exhibited an extensive, hard tumoral mass with a fluted surface measuring 30x28 cm, involving the gluteal groove and extending to the perineum with compression of the penile root (Fig.1). Upon rectal examination, the tumor filled out the ischiorectal cavity, with compression of the

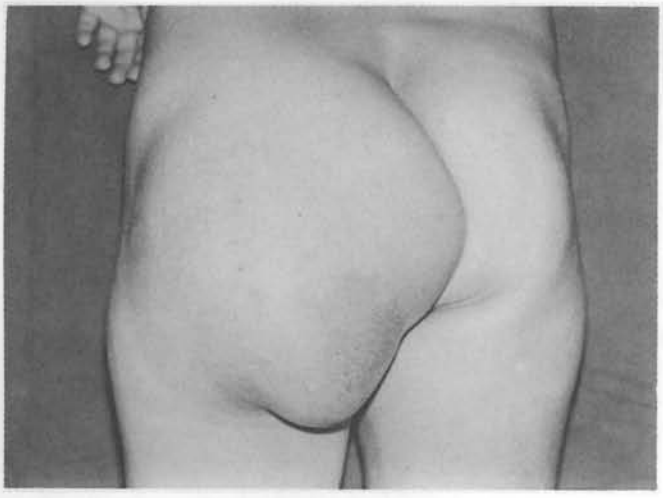

FIGURE 1 - Locally advanced chondrosarcoma.

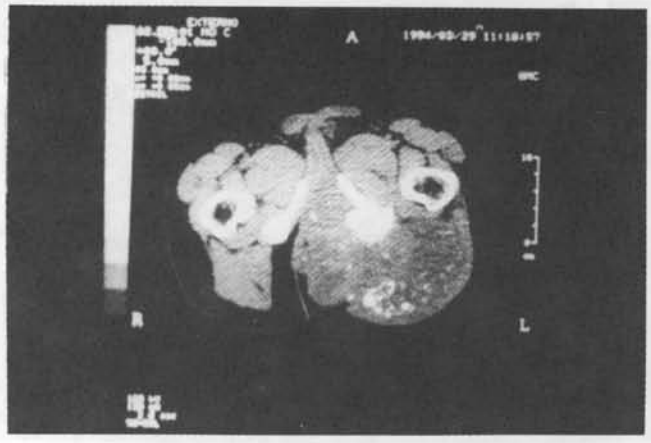

FIGURE 2 - TC scan showing tumor with calcifications destroying ischium and pubis, with extension to soft tissue.

left lateral wall of the rectum. The lesion involved bone and was attached to the skin near the gluteal groove at the most prominent part of the tumor, and at the level of the previous surgical scar.

A CT scan showed a large tumor with interior calcifications located in the left gluteal region, destroying the ischium and the pubis and extending to the acetabular region with extensive soft tissue involvement (Fig. 2). The remaining physical examination showed negative results. The remaining laboratory and clinical examinations, including hemogram, biochemical tests, chest CT, abdominal ultrasonography and ECG were normal.

A family history revealed that the patient's father, three out of four brothers, and one sister presented multiple bone exostosis, as did his two children, a girl and a boy (Fig. 3). On April 27, 1994, the patient was submitted to an extended internal Enneking[3] type II and III hemipelvectomy, the radical removal of the entire tumor, including the attached skin and surrounding soft tissue. The ischial and iliopubic branches and the acetabulum with the femur's head and neck were also removed (Fig. 4). No prosthesis was used for restoration, but a metal thread was used to attach the femur to the remaining iliac bone (Fig.5).

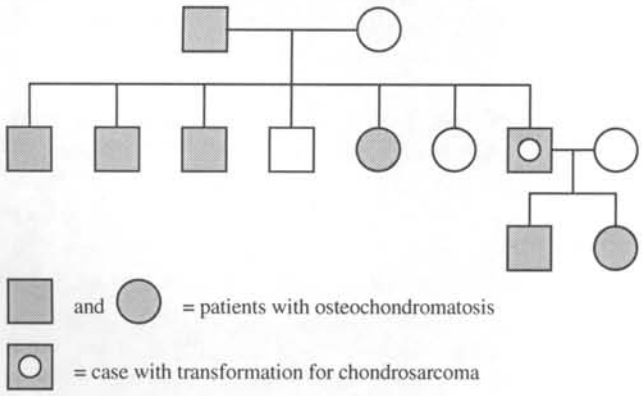

FIGURE 3 - Hereditary history pointing out the patient with transformation of the osteochondroma.

FIGURE 4 - Surgical specimen resulting from extended resection.

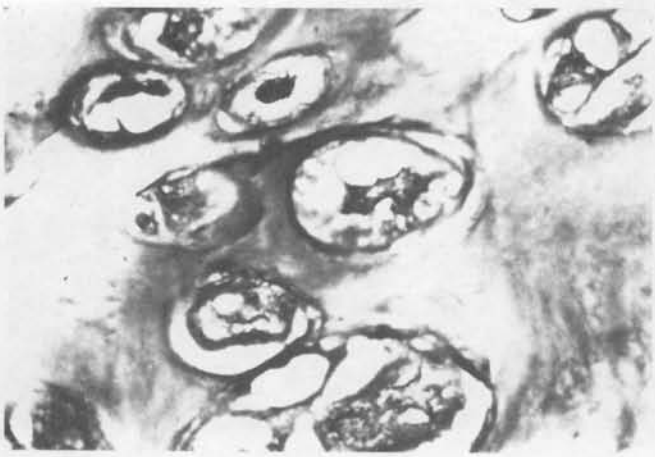

FIGURE 5 - X-ray showing loss of bone mass, metal thread attaching the femur to the iliac bone and ilio-femoral pseudo arthrosis as a result of fibrosis.

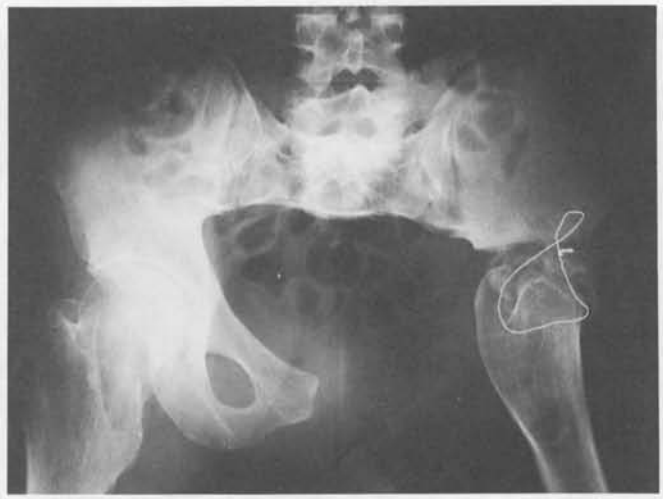

FIGURE 6 - Condrosarcoma with cell variation in shape, size, binucleation and atypical (HE - 400x).

The patient had a good postoperative recovery and started deambulation with crutches 15 days after surgery, being discharged on the day 19.

The surgical specimen measured 26x24x24 cm, including skin, muscles and bone structure of the acetabular cavity and head and neck of the femur (Fig. 4). At the cut surface, the tumor presented a white color, lobulated aspect, brittle and extending from the subcutaneous tissue and to the destroyed bone. Microscopic examination disclosed a moderately differentiated chondrosarcoma (Fig. 6) involving the skin at the level of the surgical scar, adjacent muscle and bone. No neoplasia was found on the surgical margins. The patient is alive and free of disease with positive esthetic and functional results, including deambulation with support (Fig. 7).

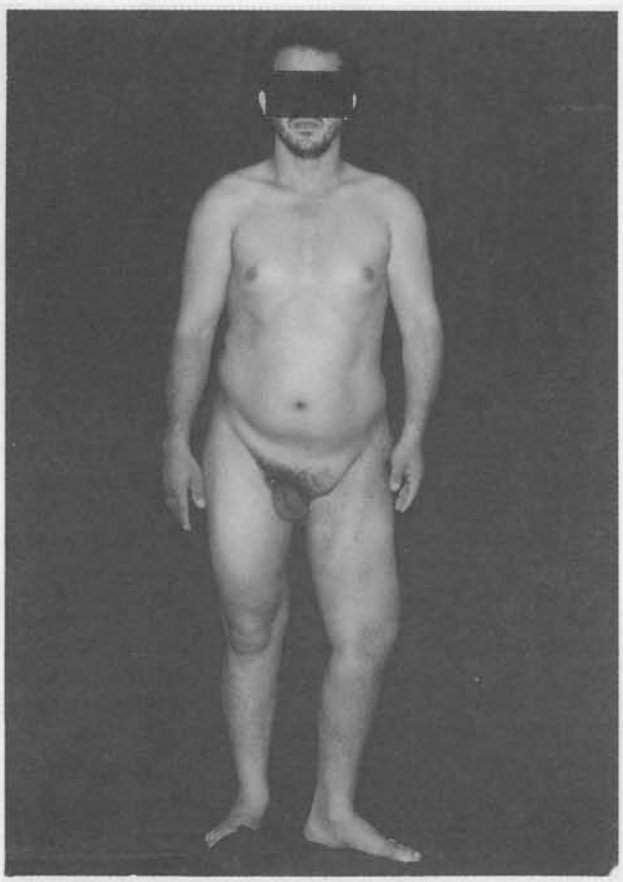

FIGURE 7 - Patient with bipedal support two years after surgery.

DISCUSSION

Secondary chondrosarcoma is related to a prior bone lesion, such as either solitary or multiple osteochondroma. Bone exostosis appears predominantly in males in early childhood, and the transformation into sarcomas occurs in adulthood, between 20-40 years.

Rapid growth, pain and calcifications are signs of malignant transformation. In HME, half of the offspring of the affected patient bear the gene and present exostosis. The hereditary history (Fig. 3) of this patient's family shows the relatives with osteochondormatosis, including this case in which malignant transformation occurred. The study of the gene involved in HME, as well as its possible mutations, might be of major significance for family planning and therapeutic counseling, as already occurs with other hereditary diseases such as familial colon polyposis.

Patients with this syndrome must be carefully followed, and any exostosis presenting signs of transformations or functional disorder resected.

Chondrosarcoma secondary to osteochondromatosis is a low-grade malignancy, and resection with adequate margins signals a good prognosis.[4]

Recently, great progress has been made in the treatment of malignant bone tumors of the pelvic girdle with limb-saving surgery procedures. Internal hemipelvectomy is little known within our surgical community. This procedure can substitute some cases of interilioabdominal amputation without impairment of the oncological margins, and improve quality of life.

In our case, an extended internal hemipelvectomy without utilization of arthrodesis or prosthesis was performed as previously described. In this case, as well as in others we have studied,[5] fibrosis formed in the stump of the amputated femur, soft tissue and/or remaining iliac bone (Fig. 7) six months after surgery. This allowed the patient nonsupported deambulation under load, and greater ease in sitting down because of the limb's flexibility around the fibrous area; this does not occur when prosthesis is used. Further, with the surgical technique used in this case, we avoided the complications due to infection frequently reported with prosthesis use.[6]

RESUMO

Os autores descrevem o caso de um paciente de 28 anos, com extenso condrossarcoma do ísquio e púbis esquerdos envolvendo articulação coxo-femural, partes moles e pele da região glútea, secundário à osteocondromatose múltipla familiar e submetido a hemipelvectomia interna tipo Enneking II e III alargada. Não se usou nenhum tipo de prótese ou artrodose. Há poucos anos, pacientes com tumores dessa extensão eram submetidos a amputação interílioabdominal com perda do membro e piora da qualidade de vida. Dois anos após a cirurgia preservadora do membro, o paciente está livre da doença, com bom resultado funcional, inclusive, com apoio bipodálico auxiliado por suporte.

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