CASE REPORT

Differential symptomatology and prevention of permanent complications dependent on localisation of osteochondroma

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ABSTRACT

Osteochondroma is the most common benign bone tumour in children and usually occurs in the metaphyseal region of the long bones, being responsible for over 40% of all bone tumours and tumour-like lesions. The objective of this paper is the presentation of a series of cases of children with osteochondroma with different localisation and clinical presentation. Variability of localisation resulted in differential approach and prevention of permanent complications. Total surgical en bloc excision of the adjacent osteochondroma is considered as the treatment of choice. Symptoms related to an osteochondroma are usually relieved by the surgery. Major complications and local recurrence are rare. The general surgical indications for benign bone growths are cosmetic defect, exostosis in a location at risk to repetitive trauma, increased risk of the exostoses to fracture, neurological involvement, impairment of the articular range of motion, and suspicion of malignancy.

KEY WORDS:
osteochondroma, exostosis, benign bone tumour, children, surgery, complications.

INTRODUCTION

The overall incidence of bone tumours and tumour-like lesions of bone is 79.3 per 1,000,000 children, as was shown in long-term population-based analysis [1]. Incidence in boys is significantly higher than in girls: 90.7 vs. 67.3, respectively. Malignant tumours (osteosarcoma, Ewing sarcoma, chondrosarcoma) constitute 17.5% of cases, and non-malignant tumours the other 82.5% [1].

Osteochondroma is the most common benign bone tumour (Table 1) and usually occurs in the metaphyseal region (Fig. 1, according to [2]) of the long bones and is responsible for 44.4% of all bone tumours and tumour-like lesions [1], although their true incidence is unknown because many cases remain undiagnosed [3]. This tumour takes the form of a cartilage-capped bony outgrowth on the surface of the bone. The vast majority (85%) of osteochondromas present as solitary, nonhe-
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TABLE 1. Benign bone tumours in children

| Group                      | Diseases                                                                 |
|----------------------------|--------------------------------------------------------------------------|
| Benign osteoid tumours     | osteoma, osteoid osteoma, osteoblastoma                                  |
| Benign chondroid tumours   | osteochondroma, exostosis osteocartilaginea                               |
|                            | enchondroma, chondroma periostale                                        |
|                            | chondroblastoma benignum fibrochondromyxoid                           |
| Other benign –tumours      | tumour gigantocellularis ossis, osteoclastoma fibro nonossificans        |
| Tumour-like lesions        | dysplasia polyostica fibrosa cystis ossi unilocuilis solitaria           |
|                            | cystis aneurysmatica ossis granuloma eosinophyllicum myositis ossificans  |
|                            | fibrous cortical defect                                                  |

Hereditary lesions. Approximately 15% of osteochondromas occur as multiple lesions in the context of hereditary multiple osteochondromas, a disorder that is inherited in an autosomal dominant manner [4]. Slow progressive osteochondroma growth is the rule in skeletally immature patients, at times prompting surgical excision [5].

Solitary osteochondromas have a tendency to appear in long bones, especially the femur, humerus, tibia, spine, and hip, although every part of the skeleton can be affected [4]. Unusual locations of osteochondromas may include clavicular involvement with impaired shoulder mechanics, which results in painful shoulder, while another with pubic ramus involvement leads to obstructive urination symptoms and dysuria. Rare locations include scapula, fibular head with peroneal or tibial nerve compression, ischial ramus, or rib involvement [6].

Hereditary multiple osteochondromas are caused by gene mutations located either on chromosome 8q24.11-q24.13 (EXT1) or 11p11-12 (EXT2), while non-heritable osteochondromas might present biallelic inactivation of the EXT1 locus (reviewed in [4]). Osteochondroma most commonly involves the metaphyses of the long bones, with a cartilage-capped bony projection on the external surface of a bone [6]. Osteochondromas develop in bones that are formed by endochondral ossification and result from displacement of the lateral portion of the growth plate, which then proliferates to the long axis of the bone [4]. The outer layer of the head of osteochondroma is composed of benign hyaline cartilage and is delineated peripherally by perichondrium. The cartilage has the appearance of a disorganised growth plate, which undergoes enchondral ossification and bone reconstruction [4].

The objective of this paper is the presentation of a series of cases of children with osteochondroma with different localisation and clinical presentation. Variability of localisation resulted in differential approach and prevention of complications. We describe detailed management in specific clinical situations.

CASE 1

A 16-year-old boy with painful tumour of the posterior medial part of the distal femur, with no history of trauma. Multiple exostoses of both femoral bones were present in a conventional X-ray study. Ultrasound examination revealed partially thrombosed pseudoaneurysm, 75 mm × 54 mm × 105 mm in size, of the popliteal artery secondary to distal femur osteochondroma. Preoperative angio-CT (computed tomography) showed partial flow in the pseudoaneurysm in the left popliteal region. The point of artery disruption was clearly visible (Fig. 2). The
CASE 1
A 14-year-old boy with symptoms of weakness of the right upper limb for more than one year followed by development of a 5-cm tumour in the medial portion of the proximal humerus. Pain, decreased range of movement, or paraesthesia were not reported. During surgery, stretching of the humeral vessels and median nerve was noted and corrected (Fig. 3). No regrowth or dysfunction were observed during eight months of follow-up after the operation. In pathology examination osteochondroma was diagnosed.

CASE 2
A 14-year-old boy with symptoms of weakness of the right upper limb for more than one year followed by development of a 5-cm tumour in the medial portion of the proximal humerus. Pain, decreased range of movement, or paraesthesia were not reported. During surgery, stretching of the humeral vessels and median nerve was noted and corrected (Fig. 3). No regrowth or dysfunction were observed during eight months of follow-up after the operation. In pathology examination osteochondroma was diagnosed.

CASE 3
A 13-year-old boy, admitted to the hospital for acute pain in the left hypochondrium. Due to splenomegaly, haematological and immunological diseases were excluded in differential diagnosis. Chest X-ray revealed a round 3 cm area, and CT scan confirmed the presence of a sixth rib tumour located on the internal side close to junctura cartilaginea. Additionally, a soft tissue reaction was found. The tumour was successfully radically resected with a thoracoscope-assisted segmental rib resection, and no complications occurred during six months of follow-up (Fig. 4). Histopathological examination confirmed the diagnosis of osteochondroma.

DISCUSSION
We present three cases of osteochondroma with different localisations, different symptoms, and different profiles of potential complications. Osteochondroma (exostosis) is a very common bone tumour. Complications, such as exostosis degeneration, orthopaedic complications, or nerve compression, occur in 4% of cases; however, vascular complications such as pseudoaneurysm are exceptionally rare. Pseudoaneurysm is most often located on the popliteal artery [7]. Pseudoaneurysm results from an arterial injury due to exostosis. Its mechanism of development is still unknown because it is usually asymptomatic. The vascular disease genesis is due to repeated exercises, such as knee flexion, which could chronically affect the popliteal artery and produce a pseudoaneurysm. Pain, oedema, and pulsating mass are the most frequent clinical signs of pseudoaneurysm. The treatment of vascular complication is surgical, at the same time as the bone tumour. Surgical treatment of exostosis vascular complications is recommended as an urgent procedure to prevent the occurrence of irreversible damage [8].

Until 2016, a total of 101 cases arterial pseudoaneurysms associated with osteochondromas were reported worldwide, with a masculine preponderancy in 86%. Popliteal artery was the most commonly injured vessel in 77%. The treatment was open surgery with vascular repair and optimal exostectomy. Arterial repair was performed with saphenous vein grafting or lateral suture. Postoperative courses were uneventful in the vast majority of cases [9, 10]. The value of the use of arteriography to confirm
the diagnosis is limited to the benefit of non-invasive radiological methods because endovascular treatment is not relevant in the setting of osteochondroma-induced arterial pseudoaneurysm [9].

Nerve compressions due to osteochondromas are extremely rare. Immediate treatment is mandatory to regain the best possible recovery, thus all patients with peripheral nerve compression due to an osteochondroma should undergo surgery. Preoperative electromyographic examinations and radiographic evaluation, consisting of MRI and CT to provide optimal information about the lesion, are crucial. Performing nerve decompression first and en bloc resection of osteochondroma consecutively in a multidisciplinary fashion is strongly recommended to avoid peripheral nerve injury [11].

Two per cent of osteochondromas grow in the chest wall [12, 13]. On the other hand, only 10% of rib tumours are benign, and osteochondromas account for half of them [14]. These tumours typically begin to grow before puberty and continue until bone maturation is reached. Costal osteochondromas tend to grow into the chest cavity, and such lesions are rarely exophytic [14]. Most costal lesions are asymptomatic, but life threatening conditions such as pneumothorax through puncture, laceration, or friction from chest wall movement during breathing can occur [12, 13]. The consequent change in intrapleural pressure and possible lung collapse can cause an emergency medical situation.

CONCLUSIONS

In conclusion, symptoms related to osteochondroma are usually relieved by surgery. Major complications and local recurrence are rare. The general surgical indications for benign bone growths are cosmetic defect, exostosis in a location at risk of repetitive trauma, increased risk of the exostoses to fracture, neurological involvement, impairment of the articular range of motion, and suspicion of malignancy. Total surgical en bloc excision of the adjacent osteochondroma is considered as the treatment of choice [11, 15].

DISCLOSURE

The authors declare no conflict of interest.

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