The treatment of eosinophilic granuloma of the humerus with nonvascularized fibular graft and elastic nail
Mithat Oner, Emre Yurdakul and Ahmet Guney

Eosinophilic granuloma is most common in children. In this paper we describe two children with a history of local swelling and pain in the humeral area who showed pathological fracture of the humerus. Needle biopsies confirmed the diagnosis of eosinophilic granuloma. Surgical procedures were performed in both patients. Both patients showed total remission after wide resection combined with segmental nonvascularized fibular graft and elastic nail. Both patients are currently free of disease after 4-year follow-up. There are several treatment modalities in eosinophilic granulomas such as radiotherapy, chemotherapy, local or systemic steroids, curettage, bone grafting and internal fixation. Although good results have been reported with nonsurgical treatment, surgery is a more effective treatment option in selected cases. In this paper we describe two children with massive solitary eosinophilic granuloma of the humerus who were successfully treated with segment resection and fibular bloc graft. J Pediatr Orthop B 22:388–391 @ 2013 Wolters Kluwer Health | Lippincott Williams & Wilkins.

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Introduction
Histiocytosis X or Langerhans cell histiocytosis (LCH) is a neoplastic proliferation of Langerhans cells of unknown aetiology. The male-to-female ratio is 2:1. It is frequently seen in long bones, pelvis, costa and vertebral bodies. It is solitary in most cases, but it can also affect multiple sites. Classic appearance of the disease is a ‘punched-out’ lytic lesion. It is a great mimicker of osteomyelitis, Ewing sarcoma and leukaemia [1]. Eosinophilic granuloma (EG) was first described by Lichtenstein [2]. The aetiology is unknown and the course of the disease is unpredictable. It develops through the accumulation of the eosinophils and histiocytes on the medulary layer of the bone [1]. EG is the least severe and localized form of all LCH and possesses the best prognostic result. Surgical excision, radiotherapy and chemotherapy, and local or systemic steroids, either alone or in combination, are the main treatment options. Surgical treatment options are especially good for the patients with pain, with large lesions, with pathological fracture or with weakened cortex [3–5]. In this paper we describe two children with massive solitary EG of the humerus who were successfully treated with wide segmental resection with combined fibular bloc graft and intramedullary titanium elastic nail.

Case reports
Patient 1
A 5-year-old girl presented with a 4-week history of predominantly nocturnal humeral pain. Her symptoms were unresponsive to medical and physical therapy. She was previously healthy and the laboratory studies were normal. Inspection showed local swelling at the right arm and the range of motion was moderately limited because of pain. We found no local heat or tenderness. Radiographic findings showed a permeative diaphysial lesion in her right arm that destroyed the cortex and caused pathological fractures, with a severe periosteal reaction that suggested an Ewing sarcoma or osteomyelitis (Fig. 1a). Bone scintigraphy confirmed that there were no other lesions. Needle biopsy showed the cells which have characteristic coffee bean-shaped nucleus. Tissue cultures were negative. Considering the patient’s age, her increasing complaints and the size of the lesion, and because of the existence of the pathological fracture, surgical treatment was thought to be appropriate. Surgery was performed to make the diagnosis and to remove the tumour. The thin-walled tumour was resected en bloc and the defect was replaced by a 6-cm segment from the ipsilateral fibula and fixed by elastic nail across the osteotomy sites (Fig. 1b). For 4 weeks postoperatively, she used a Sarmiento brace while making passive shoulder and elbow motion movements. After 8 weeks she began active motion movement exercises. After 12 weeks, the elastic nail was removed, and the patient was allowed to make a gradual range of joint movement. Radiological follow-ups in the first year and then yearly showed complete union with complete graft incorporation and consolidation (Fig. 2a and b). At 4-year follow-up, there was no evidence of recurrence, as determined radiologically. Moreover, the arm was fully functional, without any disability (Fig. 3a and b).

Patient 2
A 6-year-old girl was admitted with left arm pain on weight bearing and motion. The positive finding on physical examination was tenderness over the left arm.
Radiographs of the left humerus showed an osteolytic lesion with extensive cortical destruction and periosteal reaction that suggested an Ewing sarcoma or osteomyelitis. The radiograph showed pathological humerus fracture similar to that in patient 1. There was neither calcification nor ossification within the lesion (Fig. 4a). All blood studies were within the normal limits. The remaining radiographic skeletal survey was normal. A fluoroscopy scan-guided biopsy was performed. Histological examination revealed the morphological condition,
which was moderately rich in Langerhans cells. Because of the patient’s left arm pain and the existence of the pathological fracture, surgery was performed to make the correct diagnosis and to remove the tumour. The thin-walled tumour was resected en bloc and the defect was replaced by an 8-cm segment from the ipsilateral fibula, which was fixed by elastic nail across the osteotomy sites (Fig. 4b). The histological examination of the surgical specimen revealed a solitary EG of the bone. For 4 weeks postoperatively, she used a Sarmiento brace while making passive shoulder and elbow motion movements. After 8 weeks she began active motion movement exercises and

(a, b) At 4-year follow-up, the arm was fully functional.
at the end of 12 weeks the elastic nail was removed. Radiological follow-up every 3 months in the first year and then yearly showed complete union with complete graft incorporation and consolidation (Fig. 5). At 4-year follow-up the child is currently free of the disease and is considered to be completely cured with total disappearance of the humeral lesion.

Discussion
Eosinophilic granuloma is one of the rarest bone tumours, representing less than 1% of tumours. In 90% of the reported cases it appears in children under the age of 10 years. There is a certain predilection to males, with a ratio of 2:1 [6]. Interestingly, both of our patients were girls. According to a study by Monroc et al. [7], the disease usually presents as a monostotic lesion affecting flat and long bones with a probability of 70%, while it might also affect the skull and the vertebral spine. A unique cell, the Langerhans cell, is diagnostic [8]. It also contains Birbeck granules, whose role is yet unknown. Eosinophils, lymphocytes, fibroblasts and foam cells may be also found in the lesion, but none of these are pathognomonic. EG can be asymptomatic or present as a local swelling, pain or tenderness. If it is large and extensive enough, it may cause pathological fractures, as in our cases. Depending on the location of the tumour, it may induce neurologic and systemic symptoms such as numbness, limping, fracture, loosening of teeth, otitis media [1] or exophthalmos. No fever or other signs of inflammation have been reported in the literature [1,2,6]. Blood tests show an elevation of leucocytes and eosinophils in ~7% of the cases. The erythrocyte sedimentation rate is above normal levels [3]. Our cases showed normal blood tests. In view of the possibility of spontaneous regression, less invasive forms of treatment with lower rates of complication are desirable. Different therapeutic approaches have been proposed for the treatment of localized LCH. In some cases local injection of corticosteroids has been described [3,8]. Surgery is reserved for patients with instability, pathologic fracture or neurologic deficit, and chemotherapy is reserved for systemic involvement. The use of chemotherapy and oral corticosteroids alone or combined is indicated in systemic disease, but rarely in solitary lesions due to the unpredictable results obtained and the toxic and oncogenic risk [4]. Conventionally, surgical therapy with segment resection and nonvascularized fibular grafting has been performed for patients with EG and good clinical results have been reported [2,5]. In our cases, we preferred the option of surgery because of the existence of pathological fractures and the large size of the lesions. We applied wide resection and segmental fibular graft with titanium elastic nail and the 4-year follow-up results were excellent.

In conclusion, surgery might be preferred as a treatment option in selected cases.

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Conflicts of interest
There are no conflicts of interest.

References