Child's Nervous System (2019) 35:1419–1422
https://doi.org/10.1007/s00381-019-04208-x

CASE REPORT

A rare case: sclerosing osteomyelitis of the frontal bone

Cihan Adanas [1,2] · Sezai Özkan [1,2] · Abdurrahman Aycan [3] · Remzi Erten [1,4]

Received: 28 February 2018 / Accepted: 21 May 2018 / Published online: 26 May 2019

Abstract

The patient presented in this study had a form of chronic sclerosing osteomyelitis (CSO) that is rarely reported in calvarial bones and has never been reported in the frontal bone in the literature. We aimed to contribute to the literature with this case study. In this study, we report a 14-year-old girl who presented with swelling and pain in the frontal bone and underwent treatment due to CSO. The patient had no history of trauma and chronic infection. We conclude that CSO should be considered in the differential diagnosis of the patients presenting with cranial swelling whose diagnosis cannot be established based on the radiological findings.

Keywords Chronic sclerosing osteomyelitis · Calvarial bones · Frontal bone

Introduction

Chronic sclerosing osteomyelitis (CSO), first defined by Garré in 1893, is a chronic disease characterized by localized bone thickening and reactive bone formation. CSO is also named nonsuppurative ossified osteomyelitis and Garré's osteomyelitis [1] and is a rare disease which is difficult to diagnose. Although the exact etiology of CSO remains unclear, it has often been associated with anaerobic infections and low-virulence microorganisms. CSO mostly affects children and young adults [2]. CSO has been well defined by maxillofacial surgeons and often occurs following dental treatments [3]. CSO mostly affects maxillofacial bones, particularly mandible. In orthopedic surgery, however, it mainly occurs in the long bones such as sternum and clavicle [4]. The osteomyelitis in such cases, as opposed to the normal osteomyelitis, is a different form of chronic osteomyelitis characterized by bone thickening and expansion with no abscess, fistula, or sequestration. Most common presenting complaint in patients with CSO is swelling and tenderness in the bone located in the affected region [5]. Histopathological examination typically reveals nonspecific signs of osteomyelitis. Bacterial cultures are often negative, and the most common laboratory finding is slightly elevated erythrocyte sedimentation rate (ESR). Mainstay treatment of CSO includes creation of a gutter, muscle flap transposition, total excision of the affected bone, hyperbaric oxygen therapy, and long-term antibiotic therapy [6].

Case report

A 14-year-old girl presented to the neurosurgery department with a swelling, pain, and tenderness in the frontal region. Physical examination revealed a 4 × 4 cm palpable mass in the left frontal bone. The patient had no history of trauma, chronic infection, and surgery. Laboratory parameters showed a slightly elevated ESR with normal CRP and white blood cell count (WBC). Serological examinations were normal. Magnetic resonance imaging (MRI) visualized a 17 × 27 × 40 mm well-defined intramedullary solid mass in the left frontal bone, showing homogenous contrast enhancement following intravenous contrast administration, isointense on T1A,

✉ Abdurrahman Aycan
abdurrahmanaycan07@gmail.com

[1] Van, Turkey

[2] Department of Orthopaedics and Traumatology, Yüzüncü Yıl University Faculty of Medicine , Van, Turkey

[3] Department of Neurosurgery, Faculty of Medıcıne, Yüzüncü Yıl University, Van, Turkey

[4] Department of Pathology, Faculty of Medıcıne, Yüzüncü Yıl University, Van, Turkey

and hypointense on T2A (Figs. 1 and 2). The scalp flap was elevated from the left frontal region and the mass was exposed, which was causing subperiosteal bone destruction. An intraoperative consultation was made with the Orthopedics department due to the presence of bone destruction. The prediagnosis of CSO was established based on this consultation, and the surgical procedure was continued accordingly. Craniectomy was performed using a drill and Kerrison rongeurs. Local dural invasion was observed and the invaded part was excised. Duraplasty was performed using autologous glial fascia. No bacterial growth was detected in the cultures. Histopathological examination was consistent with sclerosing osteomyelitis of the frontal bone (Fig. 3). Depending on these findings, the patient was initiated on a 6-week antibiotic course. No complication was observed during 12-month follow-up.

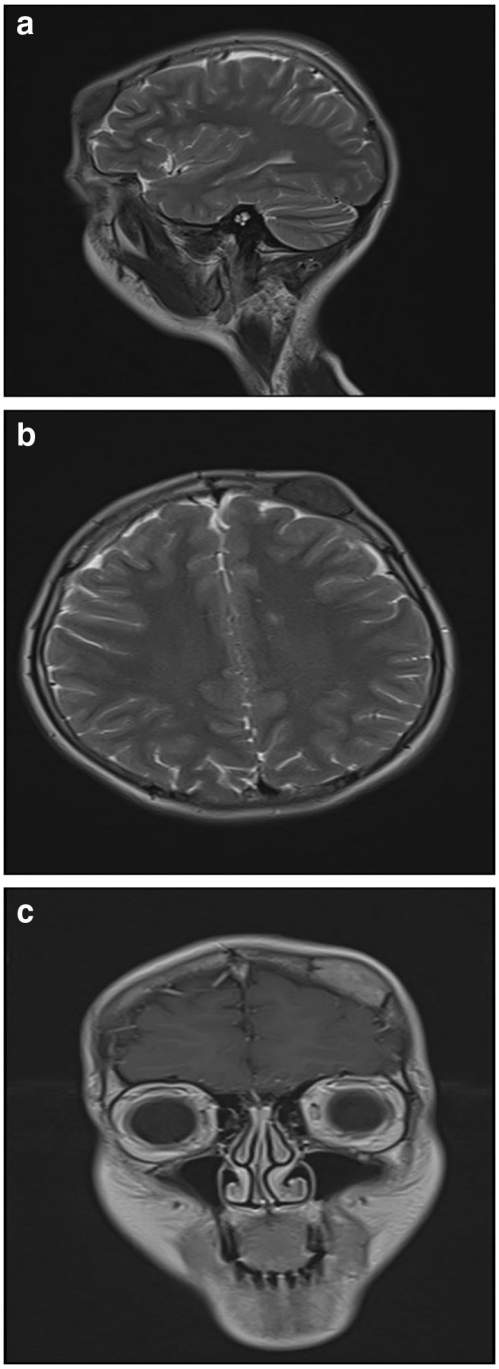

Fig. 1 Preoperative MRI images: **a** sagittal cranial image, **b** axial cranial image, and **c** coronal cranial image. The images show an impression of a sclerotic mass in the left frontal bone leading to bone destruction and swelling in the extradural space

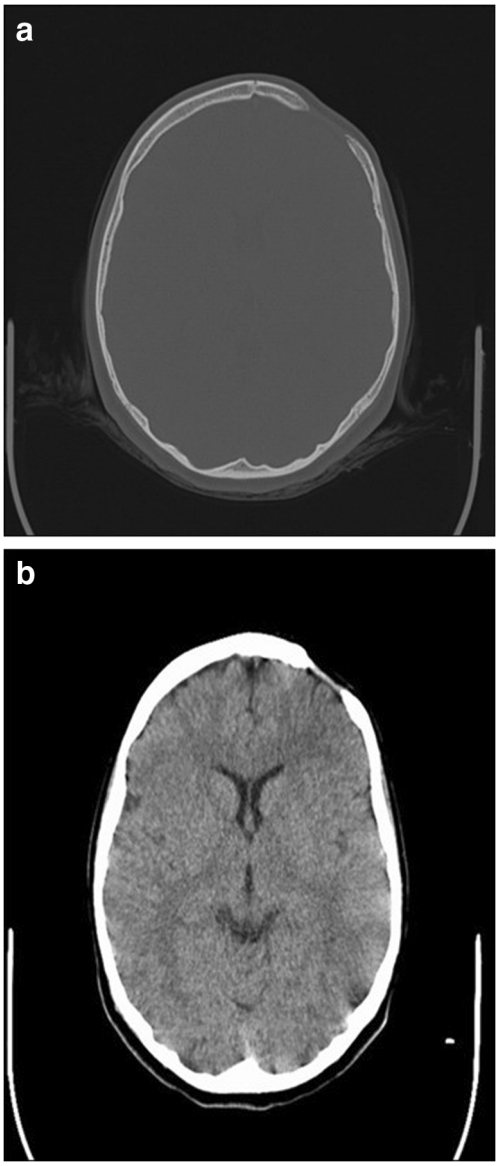

Fig. 2 **a** Cranial CT image showing bone window. **b** Cranial CT image at postoperative month 12 showing total excision of the sclerotic mass in the frontal bone and a craniectomy defect in the lesion site

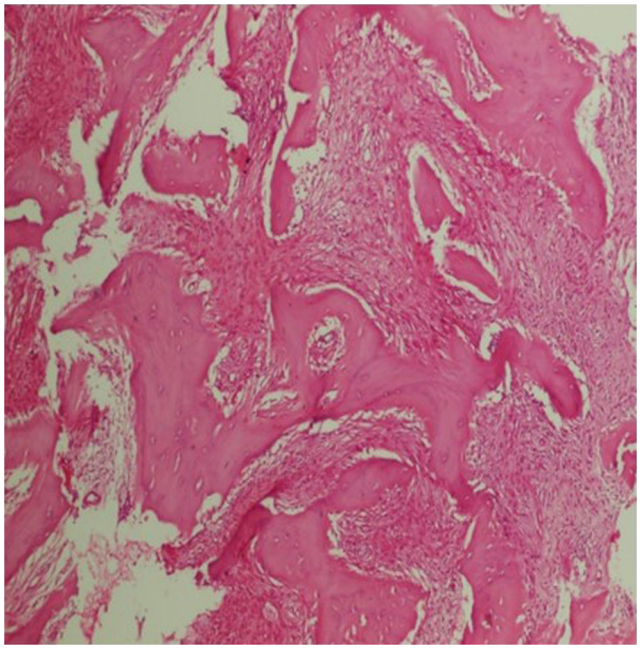

Fig. 3 Histopathological analysis of chronic sclerosing osteomyelitis showing bone marrow parenchyma replaced by the connective tissue including chronic inflammatory cells and also showing thickened trabeculae (hematoxylin-eosin × 100)

Discussion

CSO is a rare form of chronic osteomyelitis which typically remains resistant to treatment. CSO was first described by Garré in 1893 as a nonpurulent form of osteomyelitis accompanied by bone sclerosis leading to bone thickening and expansion [7, 8]. CSO often affects the maxillofacial bones, particularly mandible and maxilla, and rarely affects long bones such as clavicle, femur, tibia, fibula, and humerus [9, 10]. Literature reviews indicate that there has been only one case of calvarial CSO in the literature, which was reported by Klisch et al. [11]. To our knowledge, the case presented in our study is the second case with calvarial CSO and the first case presenting with CSO of the frontal bone in the literature.

CSO often affects children and young adults. CSO is considered to be associated with a history of trauma, chronic infection in the surrounding areas, and some low-virulence anaerobic microorganisms [2]. In patients with CSO, bacterial cultures are often negative although the growth of *Propionibacterium acnes* is rarely seen. In our patient, no bacterial growth was detected in the cultures.

Differential diagnosis of CSO often includes osteoid osteoma, intraosseous lipoma, metabolic diseases, Langerhans cell histiocytosis, intraosseous hyperostotic meningioma, osteoblastic metastases (neuroblastoma), stress fracture, fibrous dysplasia, osteosarcoma, osteoblastoma, Ewing's sarcoma, and Paget's disease.

SAPHO (synovitis acne pustulosis hyperostosis osteitis) syndrome or chronic recurrent multifocal osteomyelitis (CRMO) especially a report on calvarial lesion in the context of SAPHO was described in the literature and was recommended to be included in the differential diagnosis or sclerotic or lytic skull lesions [12]. Additionally, osteomyelitis has been reported in healthy children with frontal bones without prior trauma or infection [13].

In particular, CSO is most similar to fibrous dysplasia [2, 7–11]. On the other hand, radiological differential diagnosis of CSO is difficult and thus histopathological confirmation is mandatory. Similarly, we also performed histopathological examination for the tumor since radiological differential diagnosis was not feasible.

Patients with CSO can be culture-negative despite presenting with clinical complaints. However, increased ESR can be observed [7]. Common radiological findings include bone thickening and expansion. Moreover, periosteal bone formation with diffuse sclerotic changes can also be seen [5].

Histopathological findings of CSO typically include signs of low-grade osteomyelitis along with sclerotic bone formation suggestive of Paget's disease [9, 14, 15]. Sclerosis is considered to arise from decreased bone resorption and increased osteoblastic activity [11]. Some previous studies indicated that CSO is not associated with infection since the use antibiotics led to no relief of symptoms and thus CSO can be associated with reactive bone deposition [15].

There is still no consensus on a standard treatment method for CSO. Although some studies reported that wide-spectrum antibiotics resulted in temporary pain relief [16], some other studies suggested that antibiotics have no effect on pain relief [13, 17]. On the other hand, Jones et al. reported that calcitonin and bisphosphonate led to a significant improvement in the symptoms of pain and swelling when long-term antibiotics and surgical debridement failed to resolve these symptoms. The authors noted that the symptoms resolved completely within 2 weeks [18]. In our patient, a 6-week antibiotic course was administered and no additional treatment was required.

Surgical options for the treatment of CSO include total excision of the affected bone, bone window creation, and intramedullary reaming [19]. Lok et al. performed creation of a gutter and muscle flap transposition for the treatment of the patients with CSO of long bones. The authors noted that the use of these techniques resulted in increased blood supply and significant improvements with no need for secondary interventions [6]. In our patient, the affected bone was removed totally and duraplasty was performed for local dural invasion.

Conclusion

CSO should be considered in the differential diagnosis of all the calvarial bone tumors abovementioned. Although CSO is difficult to diagnose since radiological diagnosis is difficult and the patients often present with normal laboratory values,

Garré's osteomyelitis should be kept in mind in the diagnosis of CSO. In the CSO cases confirmed by histopathology, wide-spectrum antibiotics should be used to prevent recurrence.

Limitation

Lateral cranial X-ray images of the patient were not available in our database.

Compliance with ethical standards

Conflict of interest On behalf of all authors, the corresponding author states that there is no conflict of interest.

References

1. Wood RE, Nortjé CJ, Grotepass F, Schmidt S, Harris AM (1988) Periostitis ossificans versus Garré's osteomyelitis. Part I. What did Garré really say? Oral Surg Oral Med Oral Pathol 65(6):773–777
2. Dabov GD (2008) Osteomyelitis. In: Canale ST, Beaty JH (eds) Campbell's operative orthopaedics. Chapter 16, 11th edn. Mosby, Philadelphia, pp 695–722
3. Oulis C, Berdousis E, Vadiakas G, Goumenos G (2000) Garré's osteomyelitis of an unusual origin in a 8-year-old child. A case report. Int J Paediatr Dent 10:240–244
4. Küçükkolbaşı H, Esen A, Ataoğlu H (2007) Periostitis ossifikans (garre osteomyeliti): İki olgu sunumu. Selcuk Dental Journal 16: 42–45
5. Nasir N, Aquilina K, Ryder DQ, Marks CJ, Keohane C (2006) Garré's chronic diffuse sclerosing osteomyelitis of the sacrum: a rare condition mimicking malignancy. Br J Neurosurg 20:415–419
6. Lök V, Bal E, Sebik A, Aydinok H (2009) Long-term results of treatment including creation of a gutter and muscle transposition for chronic sclerosing osteomyelitis. Acta Orthop Traumatol Turc 43(6):510–514
7. Segev E, Hayek S, Lokiec F, Ezra E, Issakov J, ub W (2001) Primary chronic sclerosing (Garre's) osteomyelitis in children. J Pediatr Orthop B 10:360–364
8. Kelkar AS, Malshikare VA (2005) Chronic sclerosing osteomyelitis of a metacarpal. J Hand Surg (Br) 30:298–301
9. Féry A (1990) Chronic sclerosing osteomyelitis. Apropos of 4 cases and the value of the treatment by closed intramedullary reaming. J Chir 127:157–163
10. Mollan RA, Craig BF, Biggart JD (1984) Chronic sclerosing osteomyelitis. An unusual case. J Bone Joint Surg (Br) 66:583–585
11. Klisch J, Spreer J, Bötefür I, Gellrich NC, Adler CP, Zentner J, Schumacher M (2002) Calvarial sclerosing osteomyelitis. Pediatr Neurosurg 36(3):128–132
12. Dimeco F, Clatterbuck RE, Li KW, McCarthy EF, Olivi A (2000) Synovitis, acne, pustulosis, hyperostosis, and osteitis syndrome presenting as a primary calvarial lesion: case report and review of the literature. J Neurosurg 93(4):693–697
13. Arnold PM, Govindan S, Anderson KK (2009) Spontaneous cranial osteomyelitis in an otherwise healthy ten-year-old male. Pediatr Neurosurg 45(6):407–409
14. Collert S, Isacson J (1982) Chronic sclerosing osteomyelitis (Garré). Clin Orthop Relat Res 164:136–140
15. Donnelly S, Doyle DV (1993) Chronic diffuse sclerosing osteomyelitis of the humerus: novel treatment with calcitonin. J Rheumatol 20:1073–1076
16. Vienne P, Exner GU (1997) Garré sclerosing osteomyelitis. Orthopade 26:902–907 [Abstract]
17. Wright SA, Millar AM, Coward SM, Finch MB (2005) Chronic diffuse sclerosing osteomyelitis treated with risedronate. J Rheumatol 32:1376–1378
18. Jones J, Amess TR, Robinson PD (2005) Treatment of chronic sclerosing osteomyelitis of the mandible with calcitonin: a report of two cases. Br J Oral Maxillofac Surg 43:173–176
19. Bettin D, Rolauffs B, von Eiff C, Sciuk J (1999) Reconstructive treatment of sclerosing osteomyelitis of the entire femur of 30 years' duration with avoidance of segmental resection. Arch Orthop Trauma Surg 119:228–231