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Primary Chornic Osteomielitis of the Jaw in Childhood: A Case Report and Review of the Literature

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Abstract

Introduction: Primary chronic osteomyelitis is a nonbacterial inflammation of the bones; in the maxillofacial region mainly appears in the jaw. The treatment of this disease may require both surgery and anti-inflammatory drugs for a long time.

Case report: We introduce the case of a 12-year-old boy with local jaw swelling for some weeks. The patient had a growth deficit treated with growth hormone. A facial CT scan showed an alteration in the bone structure of the left mandibular ramus. The patient was undergone to surgery to obtain a bone sample diagnosing of PCO. The patient was treated with a decortication of the body of the jaw and long-term anti-inflammatory drugs, including oral corticosteroids for some months.

Discussion: PCO is a chronic disease that require a long-term treatment with anti-inflammatory drugs, sometimes even oral corticosteroids. Furthermore, decortication and curettage of the mandible region involve seem to be useful to solve the disease.

Conclusion: PCO of the jaw in children is a rare disease difficult to be diagnosed. Until now, no association has been demonstrated between PCO and the administration of growth hormone. Our results suggest that the decortication of the jaw is a good option for those cases with frequent relapses and poor control of symptoms.

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Introduction

Osteomyelitis is an inflammatory process of cortical bone that might be classified, as acute or chronic depending on the duration of the disease. Chronic osteomyelitis term is used when the evolution is longer than 4-6 weeks, from the onset of the clinical symptoms. Osteomyelitis could also be classified as primary or secondary [1]. Primary Chronic Osteomyelitis (PCO) is a nonbacterial inflammation of the bones with non-defined etiology. On the other hand, Second Chronic Osteomyelitis (SCO) is caused by bacterial infection and is more prevalent than PCO [2].

In childhood, PCO is a rare disease, which is more frequent in female patients. There is a peak at 10 years of age, with a range from 8 to 20 years [3-4]. It can be a multifocal entity or a solitary lesion. In the maxillofacial region appears mainly in the mandible [1]. The skull involvement is rarely described, most frequently in occipital bone but, if this condition appears, it must be considered in every case as a potential malignant lesion [5].

The pathogenesis of PCO is not confirmed but most experts consider, in multifocal cases, the involvement of pro-inflammatory cytokines, such as IL-6 and TNF-α, and anti-inflammatory cytokines, such as IL-10, in its progression [5,6]. In solitary mandible lesions, an alteration in blood supply to the mandible could explain isolated bone involvement [7].

Clinical manifestations are variable between patients. According to some studies, pain may be the first symptom, followed by local swelling in the affected region [1-3]. Furthermore, patients may manifest limitation in mouth opening, paresthesia of the involve area, even local lymphadenopathies with no pus formation, fistulas nor sequestration. The symptoms appear randomly and with non-well-defined onset. Likewise, the symptoms are different in intensity, quality, and duration. The presence of fever is variable [4,5,8].

PCO is difficult to diagnose and may include several diseases in the differential diagnose such as infections, trauma, malignancies (osteosarcoma, Ewing sarcoma) or metabolic disorders. Usually, the diagnosis is reached by exclusion. Laboratory tests are unspecific: they can reveal a moderate increase in white blood cell count, C-Reactive Protein (CPR) and other infectious markers but they may also be normal. As previously said, PCO is a non-bacterial inflammatory disease, thus all cultures taken from blood or the involved area are negative, excluding microbiological origin. Imaging with CT scan or Magnetic Resonance Imaging (MRI) and biopsy have an important role in diagnosing PCO [5,8].

Treatment of PCO of the jaw is frequently done with anti-inflammatory drugs such as corticosteroids or Nonsteroidal Anti-Inflammatory Drugs (NAID), which can plenty improve patient's life [1], in combination with surgical remove of the affected bone [9]. In several cases, this treatment is followed by a long antibiotic cycle and, in patients who do not respond to antibiotics, immunosuppressor treatment may be useful.

Case presentation

A 12-year-old boy with no previous medial history, except a growth delay treated with growth hormone the fourteen previous months (nine months, with 0.75 mg/kg/day and five months with 0.90 mg/kg/day) referred a 20-day history of painful swelling at the left mandible angle without fever. He was evaluated by an Odontologist who established the diagnosis of dental infection, and he was treated with oral amoxicillin-clavulanic, adjusted by weight. 2 weeks later, the patient was transferred to the Oral and Maxillofacial Surgery Department of the Marqués dd Valdecilla Hospital in Santander (Spain), due to persistence of symptoms with fever (39.3 ºC) and ipsilateral submandibular lymphadenopathy. There were no signs of oral infection, caries, or periodontitis. The blood tests were normal except for slight leukocytosis (13.300 /μl) and an elevation of C-Reactive Protein (CRP) (7.8 mg/dl). Cervical echography showed a non-specific parotid inflammation. Although several conditions could fit with the differential diagnosis, unspecified mumps or odontogenic infection were the most probable. A facial CT scan showed an alteration in the bone structure of the left mandibular ramus. The patient was admitted and treated with endovenous amoxicillin clavulanic. After three days, he improved and was discharged, under oral antibiotic therapy, with diagnosis of secondary chronic osteomyelitis from a non-identify odontogenic foci.

One month later, the patient came back to our service because of facial inflammation in the left masseter region and inability to open his mouth. He had no fever or other systemic symptoms. Serum inflammation markers were normal (CRP 0.7 mg/dl and no leukocytosis). A MRI was performed showing an alteration in de medullary bone of the left mandible from last tooth to the mandibular ramus, not affecting the condyle. The signal had a T2 hyper intensive signal with periosteal reaction and enlargement of soft tissue (masseter and pterygoid muscles). After two days of endovenous amoxicillin-clavulanic and oral ibuprofen, the patient improved and was discharged with the same previous diagnosis.

Three weeks later, the patient returned to the hospital referring a left mandibular pain, with probable inflammation of the primary tooth (#75) and swelling. He had not leukocytosis, but Glomerular Sedimentation Rate (GSR) was high (60 mm/h). At the physical examination, we observed swelling in the left mandibular angle, with open mouth range limitation and pain at the percussion in the left inferior and posterior tooth. We decided to do a dental extraction of the left inferior decidual second premolar (#75), with alveolar curettage and referred to histopathologic study. It was described as inflammatory tissue without malignancy.

After two weeks with oral treatment (amoxicillin-clavulanic and ibuprofen), the patient came back for a new episode of trismus with no fever or leukocytosis and CRP in normal range. A new facial MRI was carried out showing an unspecific lesion in left mandibular ramus from retromolar trigone and without condyle affection, involving perilesional soft tissues that were enlarged. Thus, we went under surgery to do dental extraction of the left inferior second molar (#37) and alveolar curettage. The final pathological diagnosis was consistent with a dentigerous cyst.

The patient was under follow-up and kept GSR elevated for weeks, so we decided to make a gallium-67 and technetium-99m scan that showed an abnormal signal in left mandibular region. Three months later, the patient came back for a new episode of trismus and facial inflammation, which did not improve with NSAID and muscle relaxants. A CT scan (**Figure 1**) revealed in axial slices a thickening of the left ascendant ramus with periosteal reaction with some bone destruction zones and inflammation of perilesional soft tissue. Thus, with a suspicious diagnosis of chronic primary mandibular osteomyelitis, extensive decortication of the body and angle of the mandible and

additional curettage of the bone was performed (**Figure 2**). The pathologist described the sample as suggestive of chronic osteomyelitis, so we confirmed the diagnose of mandibular PCO.

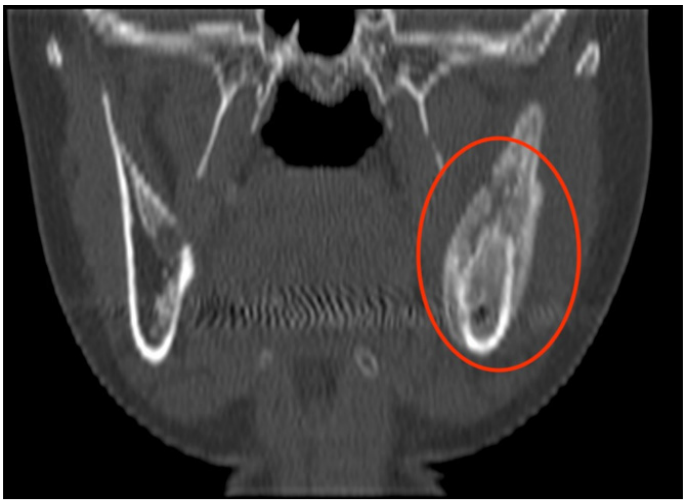

Figure 1: CT-Scan from our 12 years old patient. Circle in red, we see the ascending mandibular ramus showing bone destruction, osteolysis and perilesional tissue inflammation.

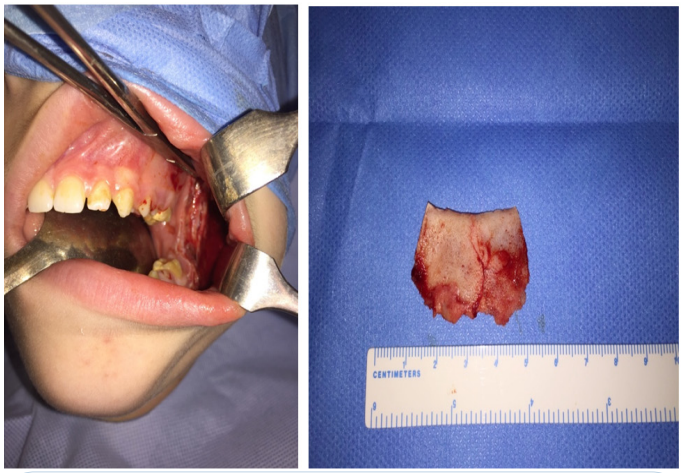

Figure 2: On the left, left mandibular decortication of the ascending ramus is observed. On the right, bone cortical sample obtained from the mandibular decortication (B).

Afterwards the patient was treated with oral amoxicillin clavulanic and prednisone (started with 10 mg per day) followed by a slow descendent pattern along next three months. Once the corticosteroids were finished three months after surgery, he began with left mandibular swelling and limited jaw range of motion so we kept low oral corticosteroids pattern for two more weeks. Nowadays, the patient is under follow-up with periodical clinical and radiological evaluation. He keeps asymptomatic for 54 months.

Discussion

Primary chronic osteomyelitis is an orphan disease which pathogenesis remains unknown. It has been suggested as an autoinflammatory/autoimmune disorder which diagnosis is for exclusion [1]. Until now, few cases with onset during childhood have been published. In fact, it has been postulated as different clinical entity instead of as merely an early onset of adult disease [10].

Although it is more frequent in girls, our case, a boy, presented the classic symptoms (pain, swelling of mandible) with several relapses as referred to in the literature [1-3]. In pediatric age, due to the nonspecific laboratory results and symptoms, the diagnosis is often difficult and often delayed in time. In this case, the absence of pus, bone sequestration or the formation of fistulas would support the diagnosis of PCO. The slightly elevated CPR and GSR values, assessed retrospectively, could suggest an inflammatory process rather than an infectious one, but without being able to be ruled out, as there is no biological marker to confirm the presence of PCO.

Despite their lack of specificity, imaging tests are the best option when it is necessary rule out other pathologies (bone tumors, dysplasias, Ewing's sarcoma, histiocytosis) but they are usually imprecise for the diagnosis of PCO. In fact, those carried out along time in our patient did not show suggestive changes in the early stages that would suggest the diagnosis. The typical appearance of PCO of the jaw on CT scan images is that the axial slices can reveal a thickening of the bone with strong periosteal reaction [4,10,11]. MRI seems to be highly sensitive in early disease; it can discover bone edema earlier than the sclerosis and osteolysis appears, helping to identify perilesional soft tissue inflammation faster than CT scan and could play and important role to identify sequelae [12].

There are few references to establish which is the best therapeutic option for this disease. The administration of antibiotics in this process is highly debated [12], and although it was applied by most authors, it also did not prove to be effective in our case, making an infectious agent in its development quite unlikely. However, as has been observed in other patients [1], the use of NIADS clearly improved symptoms but without modifying the course of the disease. Given the cyclical evolution of the disease, it is difficult to establish the impact of medical treatment on evolution [11]. Even though in general this is a relatively mild entity independent of the applied treatment, the different proposed surgical techniques are aimed to remove the necrotic tissue. For this reason, it has been suggested that decortication should be considered especially for children and adolescents [8].

Conclusion

PCO of the jaw in children is a rare disease difficult to be diagnosed. Cyclic symptoms (pain, swelling of mandible) and scarce response to usual treatments should make this entity be suspected. CT scan is the best imaging modality to diagnose it, but MRI seems to be highly sensitive in early disease. The final diagnosis of PCO should be confirmed by histopathology. Until now, no association has been demonstrated between PCO and the administration of growth hormone. Our results suggest that the decortication of the jaw is a good option for those cases with frequent relapses and poor control of symptoms.

References

1. Borzutzky A, Stern S, Reiff A, David Zurakowski, Evan A. Steinberg et al. Pediatric chronic nonbacterial osteomyelitis. Pediatrics. 2012; 130: e1190-e1997.

2. Kaiser D, Bolt I, Hofer M, Christa Relly, Gerald Berthet, et al. Chronic nonbacterial osteomyelitis in children: A retrospective multicenter study. Pediatric Rheumatology 2015; 13: 25-31.

3. Skrabl-Bumgartner A, Singer P, Greimael T, et al. Chronic nonbacterial osteomyelitis: A comparative study between children and adults. Pediatric Rheumatology. 2019; 17: 49.

4. Berglund C, Ekströmer K, Abtahi J. Primary chronic osteomye litis of the jaws in children: an update on pathophysiology, radiological findings, treatment strategies, and prospective analysis

of two cases. Case Reports in Dentistry. 2015; 2015: 12.

5. Taddio A, Ferrara G, Insalaco A, Manuela Pardeo, Massimo Gregori, et al. Dealing with Chronic Non-Bacterial Osteomyelitis: A practical approach. Pediatric Rheumatology. 2017; 15: 87.

6. Hofmann SR, Roesen-Wolff A, Hahn G, Christian M Hedrich. Update: Cytokine dysregulation in chronic nonbacterial osteomyelitis (CNO). Int J Rheumatol. 2012; 2012: 310206.

7. Bevin CR, Inwards CY, Keller EE. Surgical management of primary chronic osteomyelitis: A long-term retrospective analysis. J Oral Maxillofac Surg. 2008; 66: 2073-2085.

8. Padwa BL, Dentino K, Robson CD, Sook Bin Woo, Kyle Kurek, et al. Pediatric chronic nonbacterial osteomyelitis of the jaw: Clinical, radiographic, and histopathologic features. J Oral Maxillofac Surg. 2016; 74: 2393-2402.

9. Theologie-Lygidakis N, Schoinohoriti O, Iatrou I. Surgical management of primary chronic osteomyelitis of the jaws in children: A prospective analysis of five cases and review of the literature. Oral Maxillofacial Surgery. 2011; 15: 41-50.

10. Heggie AA, Shand JM, Aldred MJ, AA Talacko. Juvenile mandibular chronic osteomyelitis: A distinct clinical entity. Int J Oral Maxillofac Surg. 2003; 32: 459-468.

11. Eyrich GHK, Baltensperger MM, Bruder E, Graetz KW. Primary Chronic Osteomyelitis in childhood and Adolescence: A retrospective analysis of 11 cases and review of the literature, J Oral Maxillofac Surg. 2003; 61: 561-573.

12. Morbach H, Girschick HJ. Chronic non-bacterial osteomyelitis in childhood-A comprehensive review. Curr Rheumatol Rev. 2013; 9: 17.