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CASE REPORT

Total mandibular reconstruction following diffuse sclerosing osteomyelitis

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Abstract

Background Diffuse sclerosing osteomyelitis (DSO) is a non-purulent chronic recurrent inflammation and affects the mandible in many cases. Belonging to the group of autoinflammatory diseases, in children and in cases with various additional symptoms including synovitis, acne, pustulosis, hyerostosis, and osteitis (SAPHO syndrome), therapy usually consists of non-surgical treatment. Against this background, we present an unusual course of DSO in an adult female patient.

Case report A 50-year-old female suffering from DSO without SAPHO syndrome was pretreated for years with conservative drug regimens and local surgery. Previous therapy was not successful, and subsequently, multiple surgical procedures were carried out focused on recurrent acute exacerbations of DSO. Surgery resulted in a total resection and alloplastic and autoplastic reconstruction of the mandible including both temporomandibular joints. Prosthetic rehabilitation was possible after dental implant loading, and the final outcome was very satisfactory.

Conclusion In the event that non-surgical options are not successful in DSO, an extended surgical therapy becomes necessary. Even if surgery results in complete resection of the mandible, a satisfactory rehabilitation can be achieved after complex reconstruction.

Keywords Diffuse sclerosing osteomyelitis · SAPHO syndrome · Free fibula flap · Temporomandibular joint prosthesis · cad/cam · Implants

Background

Diffuse sclerosing osteomyelitis (DSO) almost takes the clinical course of a non-purulent, chronic, recurrent inflammation of the mandible. Therefore, in children and adolescents, DSO has been described as a "chronic recurrent multifocal osteomyelitis (CRMO)" and affects not only the mandible but also the long tubular bones. DSO and CRMO are known to be part of SAPHO syndrome. The acronym SAPHO describes a syndrome which includes findings of synovitis, acne, pustulosis, hyerostosis, and non-purulent osteitis [1, 2]. DSO, CRMO, and SAPHO syndrome belong to the group of inherited autoinflammatory diseases. Hence, therapy is basically conservative and consists of occlusal splint therapy; physiotherapy; application of non-steroidal anti-inflammatory drugs (NSAIDs) or, in some cases, of cortisone boosts; or even bisphosphonates [3–5]. Surgery becomes crucial only if complications arise such as pathological fractures. Against this background, we present an uncommon course of DSO in an adult female patient without SAPHO syndrome which did not respond to conservative treatment, thus demanding necessarily extended and varied surgical procedures.

Case report

A 50-year-old female was admitted to our clinical center with painful swelling, trismus, redness, excessive heat of the right cheek, and numbness of the lower lip and chin. Medical history revealed successive extractions of all teeth during the last 2 years, recurrent surgery comprising local decortications of the mandible via an intraoral approach and long-term therapy with different antibiotics, NSAIDs, and cortisone boosts. No anti-resorptive drug therapy, irradiation, or chemotherapy were applied before. A rheumatoid disease had already been

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ruled out. Lesions or efflorescences of the body skin were not apparent, and the patient had never smoked. Obviously, the patient was in a depressed mood.

An orthopantomogram and computer tomographic scans showed a widespread, extensive, and diffuse sclerosing of the mandible with no evidence of formation of an abscess or pathological fracture. Bone scintigraphy of the whole skeleton revealed a massive tracer uptake in the sense of a "hot spot" exclusively by the right body of the mandible (Fig. 1). Clinical, radiological, and nuclear medical findings led to the diagnosis of an acute exacerbation of chronic DSO. Since conservative treatment and local surgery were not successful in the past, extended surgery was planned and approved by hematology–oncology and rheumatology boards. Surgery comprised hemi-mandibulectomy including the mandibular body from the angle to the midline. In the same session, the bony defect was reconstructed using a free revascularized osteocutaneous fibula flap from the left side. The cutaneous island was placed extraorally, served as a monitor for blood supply of the osseous part of the flap, and was excised a few weeks later after complete survival of the fibula flap. The further course was uneventful, and the patient had no complaints or symptoms of osteomyelitis during follow-up visits. One and a half years later, the patient was admitted again and revealed the same symptoms and findings in imaging techniques as seen at first presentation, but now on the left side. As a consequence, the patient underwent similar surgery including left-sided hemi-mandibulectomy and reconstruction with an osteocutaneous fibula flap from the right side. After being symptom-free for 1 year, eight dental implants were loaded into the neo-mandible for prosthetic rehabilitation (Nobel Biocare GmbH, Cologne, Germany) (Fig. 2). Healing was uneventful, and after exposure of the implants, the patient received a removable denture for the mandible and a conservative full prosthesis for the maxilla. After a recurrence-free interval of 2 years, the patient again suffered from symptoms of the disease on the right side. Clinical and radiological findings revealed that the right-sided ramus and condylar process were affected, with destruction of cortical structures and multiple osteolyses. Surgery comprised the resection of the remaining mandibular part on the right side including the condyle, and reconstruction using a customized, computer-aided-designed/computer-aided-manufactured (cad/cam) alloplastic temporomandibular joint (TMJ) prosthesis consisting of a polyethylene fossa component and a titanium-coated condyle component with an extension to bridge the resected mandibular angle (Zimmerer Biomet GmbH, Freiburg i.B., Germany) (Fig. 3). Six months later,

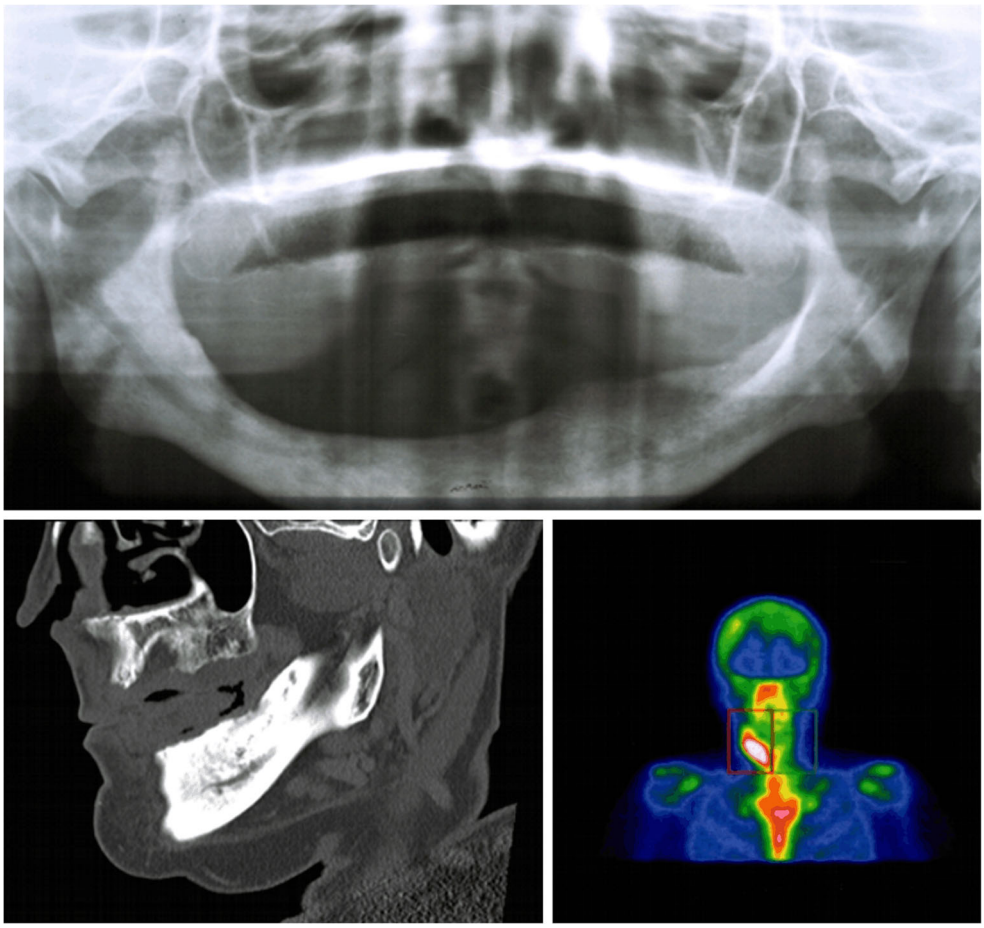

Fig. 1 Diffuse sclerosis of the mandible visible on orthopantomogram and computer tomographic scans and massive tracer uptake revealed by bone scintigraphy

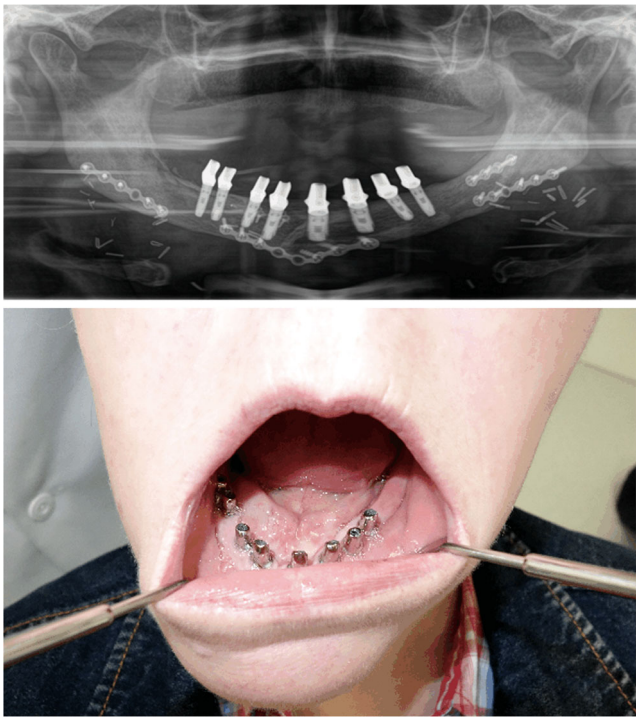

Fig. 2 Implants with abutments after reconstruction of the mandible body with two fibula flaps

similar complaints, symptoms, and findings occurred on the left side of the remaining part of the original mandible and the necessity was apparent to apply surgery to the left side in the same way as had been performed previously on the right side including a cad/cam TMJ prosthesis. Hence, the patient received an alloplastic and autoplastic, total reconstruction of the mandible (Fig. 4). During a follow-up of 1 year so far, the patient had no further complaints. All dental implants and dentures could be preserved. Occlusion was habitual, maximal interincisal opening was possible up to 30 mm, and there were no restrictions with regard to swallowing or diet (Fig. 5).

Discussion

Application of NSAIDs and cortisone boosts have been proven to be effective in cases of SAPHO syndrome, but therapy of the rare entity of stand-alone DSO is still challenging since no standard protocol exists and a complete cure is a rarity. Further conservative therapeutic "off label" options which have been considered in the literature are the application of non-nitrogen-containing bisphosphonates. However, clodronate as a representative drug of this bisphosphonate group itself has the potential to cause severe osteomyelitis and, therefore, should not be applied for standard therapy [6, 7]. There have been encouraging results from small cohort studies and anecdotal reports of use of the nitrogen-containing bisphosphonate ibandronate or the human monoclonal antibody to the receptor activator of nuclear factor kappa b ligand denusomab [8, 9]. However, well-known side effects of these anti-resorptive drugs, including osteonecrosis of the jaw, might limit their broad use in cases of DSO.

In the case presented here, DSO had previously been treated with conservative options including NSAIDs, cortisone, and local surgery without any benefit over a period of time. On the contrary, all the patient's teeth had been extracted and both inferior alveolar nerves malfunctioned. Recurrent acute exacerbations of DSO could not be controlled, and the patient developed depression and a high burden of disease. Against the background of failed conservative and local surgical therapy, the decision was taken together with members of the rheumatology and hematology–oncology boards to opt for major surgical options to achieve a visible and rapid beneficial

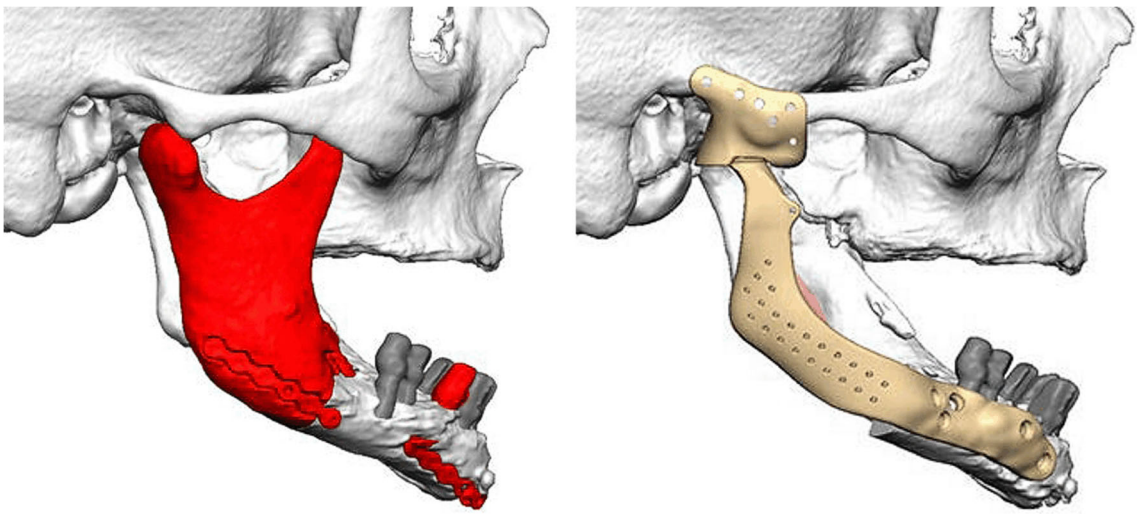

Fig. 3 Virtual planning of customized temporomandibular joint prosthesis

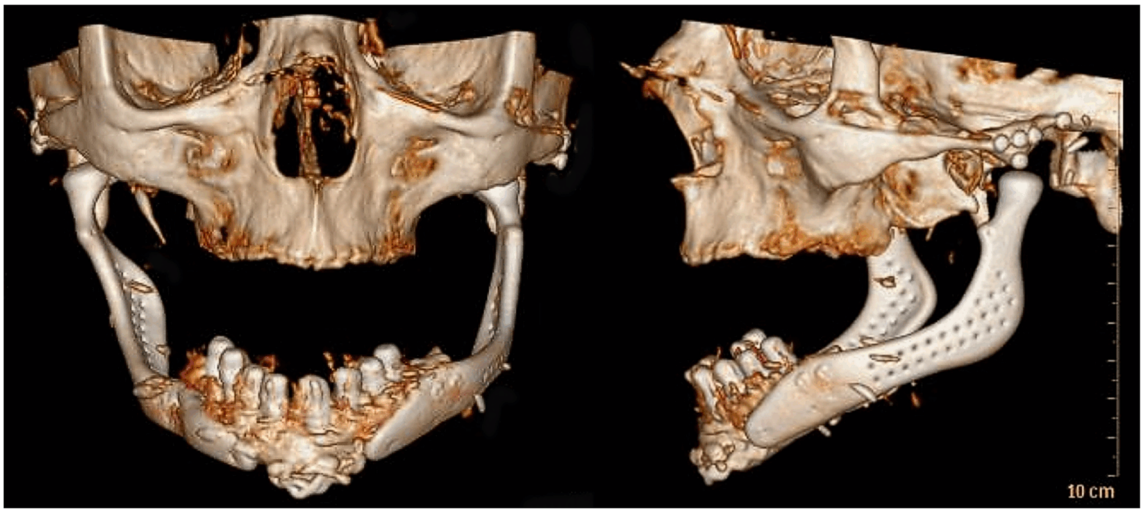

Fig. 4 A three-dimensional computer tomographic scan of the complete reconstructed mandible including both-sided temporomandibular joint prosthesis and two fibula flaps (fossa components have no radio-opacity)

outcome for the patient. Bisphosphonates or denusomab were not considered to be suitable since the impact on successful healing of a fibula flap, dental implants, or TMJ prosthesis is at least disputable. However, in the presented case, the surgery provided symptom-free intervals for many months and a comfortable outcome at the end of follow-up visits. The decision for a step-by-step procedure with partial resection and partial reconstruction of the mandible was exactly made to achieve this clinical course. Mandibular parts which were not acutely affected could be saved to hold the surgery as less extended as possible and to gain time for healing processes. Furthermore, it is not curtain that the course of DSO will inevitably result in a total mandible loss. The chance of complete healing may be small, but a long-standing chronic course without a need for further surgery would be in the scope of the disease. Hence, a complete mandible resection including not acutely affected

Fig. 5 Clinical outcome after total mandibular reconstruction

parts together with a single total mandible reconstruction in one step would be considered as an overtreatment not alone by the abovementioned board members, but also by the patient herself.

To the best of our knowledge, this is the first report of a combined alloplastic and autoplastic total reconstruction of the mandible including bilateral reconstruction of the TMJs, dental implantation, and successful prosthetic rehabilitation. Previous reports of total mandibular reconstructions did not match the term "total" exactly, since reconstructions did not include the mandibular ramus, condyle, or TMJ [10, 11]. However, in the presented case, the course of disease made extended surgery and reconstruction necessary, and the goal of total mandibular reconstruction was to achieve esthetic and functional rehabilitation.

Conclusion

Treatment of DSO in adults remains challenging, and success is inherently unpredictable. If conservative or minor surgical procedures are not successful, a major surgery with extended resections and reconstructions becomes necessary, comparable to procedures used for the treatment of malignancies. Even if surgery results in total mandibular resection, a satisfactory rehabilitation can be achieved.

Compliance with ethical standards

Conflict of interest The authors declare that they have no conflict of interest.

Informed consent Informed consent was obtained from all individual participants included in the study.

Ethical approval All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and national research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards.

References

1. Kahn MF, Hayem F, Hayem G, Grossin M (1994) Is diffuse sclerosing osteomyelitis of the mandible part of the synovitis, acne, pustulosis, hyperostosis, osteitis (SAPHO) syndrome? Analysis of seven cases. Oral Surg Oral Med Oral Pathol 78:594–598
2. Roldán JC, Terheyden H, Dunsche A, Kampen WU, Schroeder JO (2001) Acne with chronic recurrent multifocal osteomyelitis involving the mandible as part of the SAPHO syndrome: case report. Br J Oral Maxillofac Surg 39:141–144
3. Girschick HJ, Zimmer C, Klaus G, Darge K, Dick A, Morbach H (2007) Chronic recurrent multifocal osteomyelitis: what is it and how should it be treated? Nat Clin Pract Rheumatol 3:733–738
4. Hofmann SR, Kapplusch F, Girschick HJ, Morbach H, Pablik J, Ferguson PJ, Hedrich CM (2017) Chronic recurrent multifocal osteomyelitis (CRMO): presentation, pathogenesis, and treatment. Curr Osteoporos Rep 15:542–554
5. van de Meent MM, Meshkini H, Fiocco M, Wetselaar-Glas MJM, Appelman-Dijkstra NM, van Merkesteyn JPR (2017) Conservative treatment of children with chronic diffuse sclerosing osteomyelitis/tendoperiostitis of the mandible. J Craniomaxillofac Surg 45:1938–1943
6. Sugata T, Fujita Y, Myoken Y, Kiriyama T (2003) Successful management of severe facial pain in patients with diffuse sclerosing osteomyelitis (DSO) of the mandible using disodium clodronate. Int J Oral Maxillofac Surg 32:574–575
7. Senel FC, Saracoglu Tekin U, Durmus A, Bagis B (2007) Severe osteomyelitis of the mandible associated with the use of non-nitrogen-containing bisphosphonate (disodium clodronate): report of a case. J Oral Maxillofac Surg 65:562–565
8. Otto S, Troeltzsch M, Burian E, Mahaini S, Probst FA, Pautke C, Ehrenfeld M, Smolka W (2015) Ibandronate treatment of diffuse sclerosing osteomyelitis of the mandible: pain relief and insight into pathogenesis. J Craniomaxillofac Surg 43:1837–1842
9. Otto S, Burian E, Troeltzsch M, Kaeppler G, Ehrenfeld M (2018) Denosumab as a potential treatment alternative for patients suffering from diffuse sclerosing osteomyelitis of the mandible - rapid communication. J Craniomaxillofac Surg 46:534–537
10. Balaji SM (2016) Total reconstruction of mandible by transport distraction after complete resection for benign and malignant tumors. Indian J Dent Res 27:205–212
11. Abe T, Kitada H, Sato J, Yamazaki Y, Notani K, Yamaguchi T, Kitagawa Y (2011) Successful total mandibular reconstruction using a fibular graft with long-term follow-up: a case report. Asian J Oral Maxillofac Surg 23:196–200