CASE REPORT:
Successful implant rehabilitation in Erdheim-Chester Disease

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ABSTRACT

Erdheim-Chester disease is a rare multi-system disorder with oral manifestations that may result in multiple tooth extractions and edentulism. Due to the rapid and unpredictable pattern of alveolar bone loss associated with this disease, treatment with conventional dentures is very challenging and of poor prognosis. We present a case report of a patient suffering from Erdhei m-Chester disease who was treated successfully with an implant-supported over denture, overcoming the shortfalls of conventional complete dentures.

CASE REPORT

A 50 year old male presented complaining of discomfort associated with the mobility of a few teeth. To the best of the patient's knowledge he was medically fit and well.

Intra-oral examination revealed the presence of yellow fatty-like deposits in both the attached and unattached mucosae, including the hard palate. The teeth in the affected areas were mobile and exhibited periodontal pocketing. The patient was aware of the presence of these lesions for up to one year. The lesions were biopsied for histopathological analysis.

A panoramic radiograph was taken showing extensive bone loss around the affected teeth. CT scans of the maxilla and mandible revealed erosion of the buccal cortical plate of bone in the anterior aspect of the mandible and some erosive changes in the lingual cortical plate at the same level. The maxilla showed osteoporotic changes in the posterior molar area on both sides.

Blood tests revealed a raised erythrocytic sedimentation rate, indicating the presence of a systemic inflammatory process. Histology revealed an accumulation of histiocytes with frequent Touton-like giant cells, with the histiocytes having a granular eosinophilic cytoplasm. Immunohistochemical staining showed the cells to be uniformly positive for KP-1 (CD68), and negative for S100 and SMA.

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Fig. 1 - Intra-oral lesions at initial presentation 1

Fig 2. - Intra-oral lesions at initial presentation 2

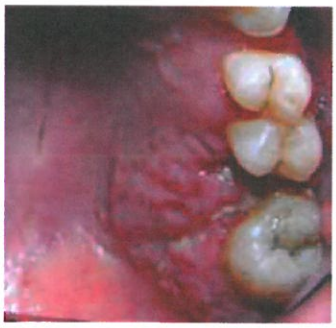

Fig. 3 - Lesion on the hard palate

Fig. 4 - Post interferon-alpha treatment 1

Fig. 5 - Post interferon-alpha treatment 2

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These findings, in combination with the reported clinical and radiologic findings, were consistent with those described for Erdheim-Chester disease.

The patient was referred for further medical investigation and management. A spiral CT scan of the chest and abdomen revealed an expansive lesion of one of the lower right ribs at the mid-axillary line, appearing to be benign.

An extensive intra-abdominal soft tissue tumour of mesenteric origin was detected just caudal to the pancreas (15cm x 8cm x 15cm) with a small amount of ascites. Bone scintigraphy showed increased tracer uptake in both tibiae, the distal halves of both femora, the maxilla, the mandible, the humeri and in the 7th and 8th ribs posteriorly.

The symmetrical uptake in the long bones was found to be in keeping with Erdheim-Chester disease. The patient was initially treated with radiotherapy to the maxilla, mandible and both tibiae. He received 1750cGy in five fractions using 6MV photons to his jaws. This treatment was of limited success.

Following this the patient was treated with daily interferon-alpha, initially 3 million units three times weekly for 9 months, increased to 6 million units six times weekly for six months and then reduced to a maintenance dose of 3 million units six times weekly. He remains on this dose to date.

During this period, the patient was regularly seen, a gradual reduction in the number and size of the lesion was noted. Eighteen months after starting the interferon, oral lesions were significantly reduced in size (fig.1-5). The patient however was still complaining of discomfort from his mobile teeth.

The first phase of treatment involved staged extractions, initially removing the lower left premolars and canine. After two months the sockets were totally healed with extensive shrinkage of the alveolus.

The remaining mandibular teeth, except for the two last standing left molars (to aid stability of the interim denture) and the mobile maxillary teeth were then removed and the patient was provided with immediate dentures. This was followed by a period of healing and observation which was uneventful other than significant remodeling leading to instability of the lower denture. Four months after the extractions, implant surgery was performed under intravenous sedation.

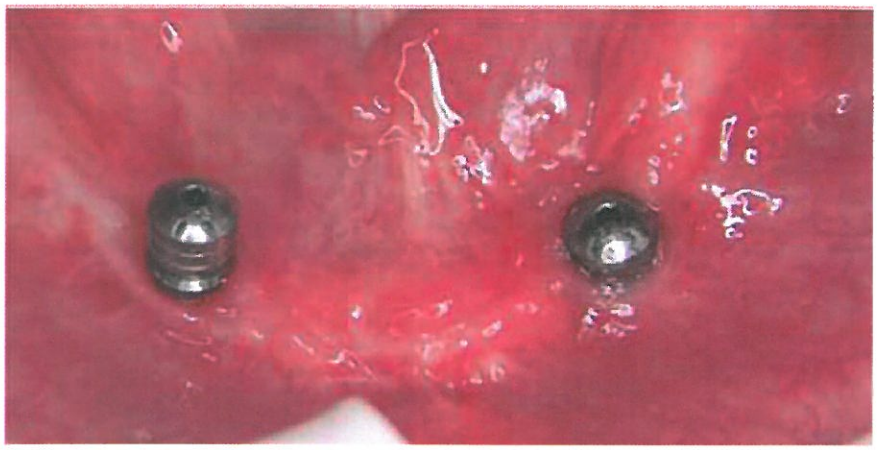

Fig. 6 - Implant healing abutments

Fig. 8 - Overdenture

Fig. 9 - End Result

Left: Fig. 7 - Gold bar

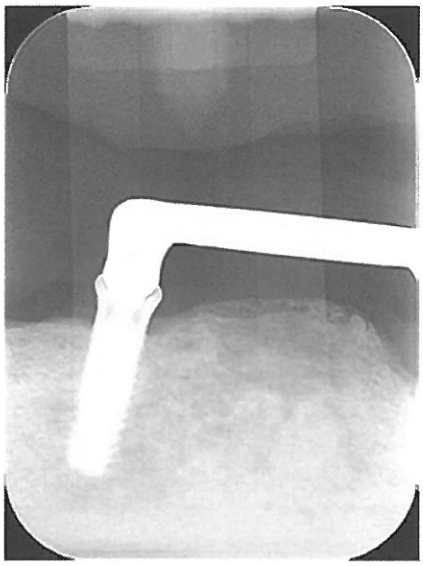

Fig. 10 - Osseointegrated Implant 1

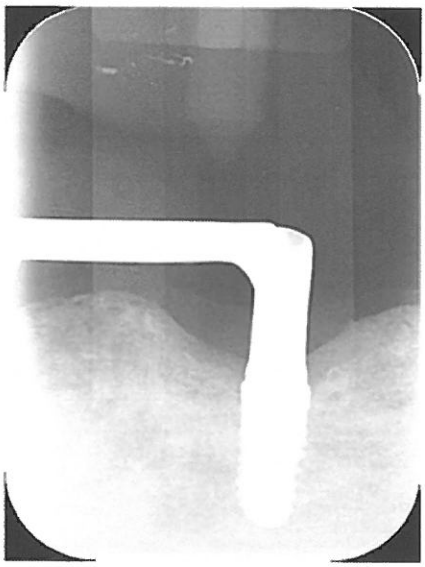

Fig. 11 - Osseointegrated Implant 2

Two implant fixtures were inserted (Dyna Dental TM implants - Dyna Dental Engineering bv, Bergen op Zoom, Netherlands) were inserted, one 3.6 x 11.5mm in the region of the lower right canine and 4.2 x 10mm in the region of the lower left canine. No abnormalities were observed in the quality of the patient's jaw bone but due to the pattern of bone loss suffered and the resulting anatomy the fixture on the left side was placed a few millimeters sub-crestally.

The initial stability was good and the torque of both implants after placement was recorded at 40Ncm-1. The lower left molars were extracted a few weeks following surgery. The patient was followed up at one week, one month and three months post-op. At this stage the remaining lower molars were removed under local anaesthesia. Healing was uneventful (fig.6) and the implants were now presumed to be osseointegrated. Fabrication of the implant-supported mandibular overdenture was carried out over the following weeks (fig. 7-9).

Fig.10-11 show peripapical radiographs of the implants taken 6 months post-implant placement showing successful osseointegration of the fixture. The patient has been reviewed 12 months post-denture insertion and is doing well.

DISCUSSION

Erdheim-Chester disease is a distinctive pathologic and radiographic entity characterised by bilateral, symmetric sclerosis of the metaphyseal regions of long bones and infiltration of foamy, lipid-laden histiocytes 1. In 1930, William Chester was the first to describe two patients presenting with a unique histiocytic disorder which he called 'lipid granulomatosis' 2. Forty two years later, Jaffe reported a similar case and named it Erdheim- Chester disease 3.

This disease has a strong male predilection and generally presents itself within the fourth and fifth decades of life1. The diagnosis is based on radiographic, histologic and immunohistochemical findings. Bilateral symmetrical osteosclerosis involving metaphyseal and diaphyseal regions typically affecting long tubular bones are the pathognomonic radiographic bone changes seen in Erdheim-Chester disease.

The distal femur, proximal tibia and fibula are the most commonly affected bones. Other bones may be affected such as the pelvis, ribs, skull, and of most relevance to the case described, the jaws. Extraosseous systemic diseases are also a feature of Erdheim-Chester disease with hypothalamic, posterior pituitary and orbital disease being the most frequent seen. The degree of visceral involvement varies greatly between cases and this has a big influence on the prognosis. One third of cases are fatal, usually due to heart failure 1.

Histology shows an infiltration of mononuclear inflammatory cells with histiocytes and occasional lymphocytes. The histiocytes are characterised by abundant pale-staining and foamy or finely granular cytoplasm, round to oval nuclei, prominent lipid vacuoles and lack the irregular configurations typical of Langerhan's cells, distinguishing the disease from Langerhan's Cell Histiocytosis (LCH).

Immunohistochemical staining is positive to CD68 and negative to CD1a and S-100. This is an important trait which further distinguished Erdheim-Chester disease from Langerhan's Cell Histiocytosis.

Treatment modalities include radiotherapy, chemotherapy, combination therapy, steroid therapy and immunotherapy. The patient presented in this case responded best to interferon-alpha. Few reports exist in the literature on the oral manifestations of Erdheim-Chester disease.

Even fewer reports exist on the oral/dental rehabilitation of patient's suffering from Erdheim-Chester disease. Aggressive periodontal disease is the most prominent intra-oral feature of the disease. Patients present with generalised gingivitis, deep periodontal pocketing, furcation involvement on molars, gingival recession, tooth mobility and multiple missing teeth.

As a result these patients require multiple dental extractions. Radiographic features of the disease may include noncorticated radiolucencies, focal lytic lesions, patchy sclerosis, marked alveolar bone loss, loss of lamina dura of the associated teeth and loss of trabeculated bone.

Valdez et al 4 describe a case where a patient suffered marked edentulous ridge resorption once the teeth were lost, affecting subsequent success of prosthodontic treatment. Six months following dental extractions the patient returned with severely atrophied alveolar ridges. Dental intervention was recommended early in the course of Erdheim-Chester disease to preserve the alveolar bone and natural dentition.

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In the case reported by Petrikowski et al 5 significant bone loss occurred despite good oral hygiene and rigorous periodontal treatment, extending to areas beyond the root apices. Dinkar et al 6 and Nagatsuka et al 7 both describe cases presenting with osteolytic lesions of the jaws in a 69 year old woman and a 13 year old girl respectively. They also suffered from cerebral and other visceral lesions, typical of Erdheim- Chester disease.

Once the teeth are extracted, what prosthodontic options exist for these patients? Typically patients were treated with complete dentures. However, due to the rapid and irregular pattern of alveolar bone loss the patients inevitably passed through a phase of repeated denture reline's only to be left with a denture which is poorly retentive, poorly functional and generally uncomfortable to wear.

Nowadays a popular solution is to provide the patient with an implant-supported prosthesis. Research by Attard et al 8 and more recently by Harris et al 9 shows that implantretained mandibular overdentures significantly increase patient satisfaction, dental function and quality of life over and above those achieved with good quality complete conventional dentures. Dental implants were considered by Valdez et al 4 but the patient's cardiac status contra-indicated elective surgery. Even so, due to the histological changes seen in the jaw, would dental implants osseointegrate successfully? To the best of the author's knowledge, only one such case was reported. Brahim et al 10 reported a case in which a 38 year old male suffering from Erdheim-Chester disease had his teeth extracted and replaced with six implants supporting a fixed mandibular denture.

It was reported that 'during surgery, it was noted that the anterior mandible was filled with deposits of yellow material resembling adipose tissue'.

Despite the abnormal bone composition six fixture were placed which were assumed to be osseointegrated and a final prosthesis was provided. Brahim et al 10 speculated that 'presumably, enough unaffected stromal cells were present to allow sufficient bone formation for osseointegration of the implant fixtures'.

The patient described in this case report exhibited similar clinical features however implant surgery was carried out once it was believed that the bony lesions had resolved. The reasons behind opting for an implant supported overdenture as opposed to an implant supported full arch fixed bridge were two-fold; primarily for ease of maintaining good oral hygiene and secondly to keep the number of implant fixtures to a minimum in the case of failure.

It was presumed that the sites chosen for implant placement were histologically normal and that osseointegration should be successful.

However, due to the lack of literature available, there was an element of uncertainty as to the success of the treatment. All post-op follow up appointments up to twelve months post-denture insertion have shown uneventful soft tissue healing and successful implant osseointegration.

ACKNOWLEDGEMENTS

Nikolai J. Attard BChD, MSc, PhD (a,b) Mark Diacono, BChD (Leeds), FDSRCPS (Glasg) (a) (a) The Dental and Implantology Unit, St. James Hospital, G. Borg Olivier Street, Malta (b) Dean, Faculty of Dental Surgery, University of Malta Medical School, Tal - Qroqq, Malta

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