Ossifying Fibroma of the Orbit

Summary

Ossifying fibroma of the head and neck is a rare tumour which most frequently involves the upper and lower jaw. It occurs during the third and fourth decade of life. The lesion is usually solitary and well circumscribed. It grows slowly, thinning the surrounding healthy bone tissue. Radiographic appearance depends on the duration of the disease. In the early stage it presents as a sharply circumscribed zone of translucency with single accumulations of calcification. During the further course, with the sharply circumscribed zone of translucency on the periphery, the centrally located accumulations of calcification become denser. The histological appearance is characterised by fibrous tissue, interwoven with bony trabeculae of lamellar bones, which are bordered with osteoblasts and scattered myxomatous stroma. Therapy is surgical and consists of total resection of the tumour, which is the only way to prevent the occurrence of a local recurrence. In this paper we present a rare case of ossifying fibroma of the orbit which involved the zygomatic bone and spread/extended via the large branch of the sphenoid up to the optic canal. The specificity of the therapy in our female patient was that in spite of contact of the tumour with the frontal base of the skull, a neurosurgical approach was not used.

Key words: ossifying fibroma, great wing of the sphenoid, therapy.

Introduction

Ossifying fibroma belongs to a group of diseases which includes cementofibroma, benign cementoblastoma and periapical fibrous dysplasia. They all arise from primitive mesenchymal cells of periodontal tissue, which have the potential of forming bone tissue, cement or, as in the majority of cases, both of these. Thus, the different spectre of the same disease should be considered rather than particular enti-
ties. For this reason diagnosis can only be made on the basis of a combination of the clinical appearance and radiographic and histopathological finding. Ossifying fibroma of the head and neck is a rare tumour which most frequently involves the upper and lower jaw, and occurs as a rule in the third and fourth decade of life. The lesion is usually solitary and of slow growth, although aggressive course of the disease has been reported, particularly in young patients and in those in whom the disease was located in the maxilla (1). The radiographic appearance depends on the duration of the disease, and thus in the early stage it presents as a sharply circumscribed translucency with occasional accumulations of calcification. In the further course of the disease, apart from the sharply circumscribed zone of translucency on the periphery, the centrally located accumulations of calcification become denser. In contrast to fibrous dysplasia, which can involve more than one bone, has unclear borders and spreads through the bone, growing within it, ossifying fibroma is as a rule monoostotic, well circumscribed and develops slowly, thinning the surrounding healthy bone tissue (2). Histologically, the ossifying fibroma is characterised by fibrous tissue interwoven with bony trabeculae of lamella bones which are bordered with osteoblasts and in places myxomatous stroma. In some places inflammatory cells can be found, gigantic cells of the foreign body type and accumulations of hemosiderin. Ossifying fibroma of the maxilla can involve the maxillary sinus, ethmoid and orbit, and in rare reported cases the base of the frontal skull cavity (3) and temporal bone (4). Therapy is surgical and consists of total resection of the tumour, which is the only way to prevent the occurrence of local recurrence. The aim of this paper was to present a rare case of ossifying fibroma of the orbit which encompassed the zygomatic bone and spread/extended via the great wing of the sphenoid up to the optic canal.

Case presentation

A female patient, MM, aged 77 years, was referred to our Department because of protrusion of bulbus (oculi?) and deformation of the right side of the face. The case history showed that the patient had been operated on 10 years earlier because of a tumour in the right temporal region, of which the patient did not have medical documents. Two years later she was again operated on in the same institution because of local recurrence. At that time osteoma was diagnosed. Six months after the second operation the patient was referred to a maxillofacial surgeon because of protrusion of the right bulbus. CT was performed for the first time which showed a well circumscribed bone tumour, which involved the lateral wall and floor of the orbit, reducing the bone mass of the lateral edge of the orbit, body of the zygomatic bone and extended into the maxillary sinus, whose volume was almost completely reduced. The tumour also encompassed the great wing of the sphenoid and extended up to the optic canal (Figure 1).

During the operation a combination of coronary, infraorbital and intraoral approach was used. After osteotomy of the zygomatic bone it was temporarily removed, enabling adequate access to the tumour located in the orbit and infrazygomatically (Figure 2).

Firstly, the tumour was removed infrazygomatically, and then after preparation of the soft tissues of the orbit also from the area of the great wing of the sphenoid, where during resection we exposed the dural of the frontal skull base. After removal of the tumour (Figure 3) osteosynthesis was performed of the previously osteotomated zygoma with mini plates, and the defect of the lateral wall and floor of the orbit was reconstructed with titan mesh which we covered with Dexon mesh (Figure 4). The postoperative course was normal. A control CT showed that the tumour was completely removed (Figure 5). Six months after the operation the patient has normal vision of the right eye without enophthalmus or double vision. The lower position of the lateral canthus will be corrected in a further second procedure (Figure 6).

Conclusion

Differential diagnosis of benign fibrous bone tumours in the area of the head and neck, in view of their mutual origin from cells of the periodontal ligament, is very often unclear and represents a serious clinical problem. The radiographic appearance
of these changes in the early phase is almost identical which obstructs differentiation of the initial phase of ossifying fibroma from fibrous dysplasia. The same is occasionally true for the histological appearance, because of which the final diagnosis is as a rule almost always based on a combination of clinical, radiographic and histological indicators (5). Fibrous dysplasia most frequently occurs in the first or second decade of life, and the greatest incidence of ossifying fibroma occurs in the third and fourth decade (1). In the case of our patient it was unusual for the disease to occur in the eighth, and probably the late seventh, decade of life, which is unique in the literature.

The fact that CT diagnostics was not used during the preoperative planning of the first two operations, performed in other institutions, suggests the likelihood of inadequate resection, indicated by the two local recurrences. The value of computerised tomography in diagnosis not only of localisation but also the type of tumour of the orbit has long been known (6) and confirmed in clinical practice. The specificity of magnetic resonance in diagnosis of ossifying fibroma remains controversial (7, 8).

We are of the opinion that resection of an ossifying fibroma and of any other tumour of the orbit should not be undertaken without previous CT diagnostics, on the basis of which an adequate procedure can be planned. Erroneous histopathological diagnosis of the osteoma after the second operation, performed in another institution, suggests inadequate histological material, i.e. most probably only the external membrane of the tumour was removed.

It is well known that an ossifying fibroma occurs most frequently in the area of the mandible, followed immediately by the maxilla, firstly of the medial and lower orbital edge and maxillary sinus. However, cases have been reported of spreading to the ethmoid, sphenoid bone and base of the frontal skull cavity (3).

Apart from our patient only one case of ossifying fibroma of the lateral edge of the orbit has been described in the literature, although without involvement of the great wing of the sphenoid (9). Imperative total resection of this locally destructive tumour can be achieved by a combined coronary, infraorbital and intraoral approach with so-called "temporary" osteotomy of the zygomatic bone, which after removal of the tumour is again fixed in its anatomic position with mini-plates. Although during the operation, because of spreading of the process to the base of the anterior skull cavity the dural was exposed, a neurosurgeon did not participate in the operation. While some authors recommend a multi-disciplinary approach in the surgical treatment of ossifying fibroma of the orbit (10) we believe that in certain, precisely preoperatively planned cases, it is unnecessary.