Recurrent Benign Giant-Cell Lesions of the Jaws with Metastasis?

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The existence of a giant-cell tumour in the jawbones has been a matter of discussion for many years. In an excellent review Jaffe, Lichtenstein and Portis (1940) tried to clarify some of the problems by defining true giant-cell tumours of bone and separating them from reparative giant-cell granulomas. Later Jaffe (1953) further discussed some of the giant-cell containing lesions of the jaws and stated that he had only seen one case of true giant-cell tumour in the jaws. Extensive reviews by Waldron (1953), Austin, Dahlin and Royer (1959), Bhaskar, Bernier and Godby (1959) and Shklar and Meyer (1961) all concluded that giant-cell tumours very seldom occur in the jaws; Bhaskar et al. even denied the existence of this tumour in the jaws. In his recent textbook, Jaffe (1958) discussed the nature and genesis of these tumours, regarded the tumour cells as being derived from primitive mesenchymal cells in the bone marrow, and pointed out that the stromal cells are the essential tumour cells and the giant-cells are products of these stromal cells; at the same time these cells are responsible for the growth of the tumour. He divided the tumour into three grades, I - II - III, I: no appreciable, II: moderate and III: pronounced atypism of the stromal cells, grade III presenting a sarcomatous stroma, frankly malignant.

Further, Jaffe stated that even tumours of grade I might be able to metastasize. In case of recurrences the tumour cells are usually more plump than the original cells.

Since we have been able to follow a case with recurrent giant-cell containing lesions in the jaws with later appearing identical lesions of the long bones for 18 years we have felt it of interest to report the clinical, radiographic and histopathological features of this patient.

Case Report

Woman, born in 1924.

History: In 1940 the patient had been admitted to a gynaecological department because of pituitary dwarfism and genital atresia. Treated with chronic gonadotrophin. The first menstruation occurred when the patient was 22 years old. The menstruations have since been normal.

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E. Hjørting-Hansen and J. Worsøe-Petersen

Present disease: In 1945 she was admitted to the E.N.T. Department because of a swelling, slowly growing for the past 6 months and located on the left side of the nose. Clinical examination revealed a bony intumescence, situated approximately in the canine fossa. The skin was movable in relation to the tumour. Radiographs of the nasal sinuses (Fig. 1) on 7 September 1945 showed a diffuse blurring of the left maxillary antrum. There were no signs of destructive lesions. The disease was interpreted as a follicular cyst, an intraoral film (Fig. 2) showed radiolucency around and between the roots of the left upper canine and the first premolar.

On 15 September 1945 the patient had a partial resection of the tumour in the left maxilla and Caldwell-Luc resection of the left maxillary antrum.

Fig. 1.
Radiograph of the nasal sinuses, 7 September 1945. A diffuse blurring of the left maxillary sinus is seen.
Recurrent Benign Giant-Cell Lesions of the Jaws

FIG. 2.
Intraoral radiograph, 7 September 1945. Irregular radiolucencies around the roots of the left upper canine and the first premolar are seen.

The cystic tumour extended to the floor of the nasal cavity down towards the upper teeth and contained spongy, yellowish-brown tissue.

After the histological report (vide infra) was received, it was decided to try radical surgery, so that total excision of the tumour in the left maxilla was performed on 25 September 1945.

From 12 December to 31 December 1945 the patient received X-ray treatment of the maxilla, delivered to two fields, 1,500 r to each.

In 1949 she was re-admitted. One month previously she had noticed that her chin was getting larger. Objective examination showed a hard, smooth swelling on the anterior aspect of the mandible corresponding to the area from second right incisor to second left premolar. Radiographs on 17 February 1949 (Fig. 3) disclosed a radiolucency in the mandible, reaching posteriorly to the first left molar and continuing anteriorly to the midline where its contours were less regular and it grew out on the anterior aspect of the mandible.

From 19 February to 3 March 1949 X-ray therapy was delivered to the mandible in two fields, 1,000 r to each field.

On follow-up, the tumour was found unchanged. On 12 May 1949 the mandibular tumour was excised.

Out-patient follow-up on 22 August 1950 showed in the right canine fossa a tumour of a nature similar to the previous ones. On 22 September 1950 excision of the tumour in the right maxilla was carried out. This tumour was situated in the canine fossa, extending to and surrounding the piriform aperture right up to the infraorbital border. Inferiorly the tumour extended to the hard palate.

Because of a recurrence the patient was re-admitted, and on 30 November 1951 a tumour of the right cheek and maxilla was excised. This tumour was of approximately the same size and location as the last one.
At follow-up in September 1962 radiographs of the mandible showed, almost throughout the mandible, irregular translucencies of varying size. In the midline there was a large osseous defect, presumably sequelae of the previous operation. Tomography of the maxillary antra showed the sequelae of resection of the left antrum, but there were no signs of recurrence.

In 1963, however, she was again admitted. Three months before this admission she had noticed a steadily growing intumescence in the right lower labial sulcus at the site of the incisors. Clinically, there was a rounded intumescence, 2 x 5 cm., with ample vascularization on the surface. The consistency was like that of the previous tumours.

On 20 May 1963 the tumour was excised.

At follow-up on 10 September 1963 radiographic examination of the long bones was performed. This revealed mild generalized osteoporosis with cyst-like radiolucencies in the lateral humeral epicondyle on both sides (left side shown in Fig. 4). In the left head of the radius and in the right femur, at the site of the neck, similar radiolucencies were found. The distal end of the left femur contained several translucencies (Fig. 5). For comparison, Fig. 6 is a radiograph of the knee from 1945, without any translucencies. The phalangeal bones of the hands and feet also showed numerous small translucencies. The patient has refused biopsy from these areas.
Recurrent Benign Giant-Cell Lesions of the Jaws

Fig. 4.
Radiograph of the left elbow joint, 10 September 1963. Radiolucent areas are seen in the lateral epicondyle.

Laboratory examinations showed values at serum calcium 9.1 and 10.1 mg./100 ml., serum phosphorus 4.9 and 4.0 mg./100 ml., alkaline phosphates 5.2 units/100 ml. and acid phosphates 4.2 units/100 ml. Urea clearance 106 per cent. and 124 per cent.

Histological Examination

Numerous sections were examined of all biopsies and surgical specimens.

Figs. 7 and 8 represent the primary lesions of the maxilla and the mandible. A fibrous, very cellular stroma characterized the picture. Giant-cells are present, usually arranged in nodules with bundles of collagen fibres separating the nodules. The nuclei of the giant-cells are small and most frequently basophilic. Signs of fresh and old bleeding are seen diffusely through the lesional tissue.

Figs. 9 and 10 represent the recurrences in the maxilla and the mandible. A diffuse arrangement of the giant-cells is the characteristic feature.
Radiograph of the left knee, 10 September 1963. Radiolucent.
FIG. 7.
Primary lesion from the maxilla. Note nodular arrangement of the giant cells. Haematoxylin and Eosin, $\times 25$.

FIG. 8.
Higher magnification of similar area from the mandible. Haematoxylin & Eosin, $\times 100$. 

1039
FIG. 9.
Secondary lesion from the maxilla, the giant-cells are diffusely scattered through the lesional tissue. Haematoxylin & Eosin, ×64.

FIG. 10.
Higher magnification of similar area from the recurrence in the mandible. Slight variation in the stromal cells is seen. Haematoxylin & Eosin, ×400.
Recurrent Benign Giant-Cell Lesions of the Jaws

of the recurrent lesions of the jaws. The nuclei of the giant-cells resemble rather closely the stromal cells. These stromal cells are in general uniform through the lesion, although slight variations in size, chromatin pattern and staining properties of the nuclei are seen. Extravascular erythrocytes are a frequent finding.

Comments

When multiple giant-cell lesions of the nature presented here occur in the jaws or other bones several lesions have to be considered from a differential diagnostic point of view.

1. Hyperparathyroidism. This was ruled out on the basis of normal values for calcium and phosphorus in serum and a normal clearance of urea.
2. Polyostotic fibrous dysplasia. Although the histopathological picture in fibrous dysplasia varies a great deal, one would always expect to find some bone formation and very seldom the amount of bleeding as in the present case (Harris et al., 1962). Albright’s disease could also be ruled out, since none of the endocrine abnormalities characteristic for this disease were found in the present case and no abnormal pigmentation was present (Albright et al., 1937).
3. Giant-cell reparative granuloma. The histopathological picture of the primary lesions is rather characteristic for this lesion (Figs. 7 and 8). Usually, giant-cell reparative granulomas do not recur after simple curettage; in the present case the lesion in the upper jaw recurred twice and the lesion in the lower jaw once. Further, multiple occurrence of giant-cell reparative granulomas is very rare. The histopathological picture of the recurrences bears no resemblance to giant-cell reparative granuloma. The giant-cells are diffusely scattered through the lesions. The stromal cells are bigger, their nuclei resemble the nuclei of the giant-cells and occasionally slight atypism is seen (Figs. 9 and 10).

Within the follow-up period the patient developed radioluencies in the long bones. She refused to have biopsies taken. The radiographic picture was rather similar to the picture of the lesions in the jaw bones.

Bloom, et al. (1962) presented a case of an 8-year-old boy, who during the last 3 years had six giant-cell containing lesions in the jawbones and femur and radioluencies similar to the jaw lesions in the humerus and matacarpals. Three of these lesions were diagnosed as giant-cell tumours grade II according to Jaffe’s classification (Jaffe, 1958). This case is the only case the authors have found in the literature with resemblance to the present case.
E. Hjørting-Hansen and J. Worsøe-Petersen

It is interesting to note that the locations of the lesions in the long bones in the present case corresponds exactly to the most frequent location of primary single giant-cell tumours (Jaffe, 1958). The giant-cell lesions of the jaw bones presented here are difficult to classify. Further, the X-ray therapy may have caused the later aggressive nature of the lesions, and since the patient is a dwarf, disturbances in normal hormonal balance may contribute. The primary lesions both of maxilla and mandible resembled most histologically the giant-cell reparative granulomas, whereas the recurrences were more like the histological structure of a giant-cell tumour, grade I–II. The fact, that the lesions in the distal part of the femur has developed during the observation period (Figs. 5 and 6) lends support to Jaffe’s (1958) opinion that benign giant-cell tumours can metastasize.

In conclusion, the lesions in the presented case cannot be finally classified, but as mentioned earlier, the findings are similar to the case presented of Bloom et al. (1962).

Summary

A 38-year-old woman with multiple giant-cell lesions of the jaws and long bones is presented. The patient has been followed for 18 years. The primary lesions of the maxilla and the mandible resembled histologically reparative giant-cell granulomas, whereas local recurrences demonstrated a microscopical picture more like a true giant-cell tumour. Lately the patient developed similar lesions in the long bones. The nature of the lesions is discussed.

REFERENCES

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