Introduction

Desmoplastic fibroma of bone is a rare fibroblastic lesion of bone that histologically resembles the desmoid tumor of soft tissue. It’s a nonmetastasizing but locally aggressive tumour.

Clinical case

We report a case of DF of the right mandibular in a 52-year-old male treated with a wide surgical excision.

Discussion

With less than 200 cases reported in literature, DF is a very rare benign bone tumour. In the head and neck region, they are most commonly seen in the mandible and have been treated in various ways. Clinically, DF has a variable symptomatology. Radiographically, it appears as a multilocular or occasionally unilocular radiolucency with well-defined or ill-defined margins. Positive diagnosis is based on surgical pathology. The treatment consists of wide surgical excision. Radiation therapy sometimes has been used. Recurrences will depend on the quality of the treatment.

Conclusion

Remaining histology the gold standard for the DF diagnosis.

Keywords: desmoplastic fibroma, benign tumor, mandible, surgical pathology
Desmoplastic fibroma of the mandible

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A DF often behaves in an aggressive manner and macroscopically has a firm consistency with well-defined advancing surfaces that may extend into surrounding soft tissue. The histological features of DF and the extra-abdominal desmoid tumour are essentially identical. They are characterized by uniform-appearing fibroblastic cells in a stroma containing various amounts of collagen fibers. The morphologic differential diagnosis includes benign and malignant spindle cell tumours of bone. Fibrous dysplasia can stimulate desmoplastic fibroma in areas where fibrous tissue predominates and ostoid production is not apparent. The distinction can be made by recognizing areas of bone formation by additional sampling. Also the nuclei in fibrous dysplasia are shorter and more compact-looking than the elongated, slender nuclei seen in desmoplasic fibroma. Low grade intraosseous osteosarcoma, another tumour that can mimic desmoplasic fibroma, can also be excluded by identification of bone formation.

Nonossifying fibroma and solitary congenital fibromatosis of bone can be confused with desmoplastic fibroma. Low grade fibrosarcoma poses the most difficult problem in the histological differential diagnosis; in fact, the distinction may not always be possible and can only be detected when it recurs and metastasizes. However, fibrosarcoma is more cellular, with a recognizable herringbone pattern and plumper, larger cells than those in desmoplasic fibroma. Cytologically hyperchromasia with anaplasia and mitotic activity quantitatively surpasses the rare mitotic figures occasionally seen in desmoplastic fibroma.

Jaffe, in his discussion of the treatment of desmoplasic fibroma of bone, recommended segmental resection as the treatment of choice and noted that if the lesion is curetted and recurs, segmental resection or a more thorough curettage should be performed. Wide resection or a thorough “marginal” curettage was the preferred method of treatment while local or limited curettage often led to continued growth of the tumour. There are conflicting reports regarding the role of radiotherapy in the management of desmoid tumours.

In 1944, Pack and Ehrlich, stated that radiation therapy could affect regression of desmoid tumours, but this process was slow. Other authors, even recently, have judged radiation to be of limited value in the curative treatment of patients with desmoid tumours. Radiation therapy is recommended in those situations where wide-field resection without significant morbidity is not possible for gross local disease. Role of chemotherapy and hormonal therapy in the management of desmoid tumours is not clear.

Conclusion

Desmoplastic fibroma is a rare intraosseous well-differentiated benign fibrous tumour, but locally aggressive. Distinguishing DF from other spindle cell proliferations has important prognostic significance and remains a difficult process for the pathologist. Wide resection or en bloc resection with long-term follow-up continues to be the treatment of choice.

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Conflict of interest

The author declares no conflict of interest.

References


