Chapter 9
Osteofibrous Dysplasia and Adamantinoma

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These two lesions are so related as to deserve to be described under the same heading. Both are rare, but adamantinoma is three times rarer than osteofibrous dysplasia. Osteofibrous dysplasia is a benign even spontaneously regressing lesion, while adamantinoma is a low-grade malignant tumor also capable of metastasizing. There are, however, a few cases where an osteofibrous dysplasia changed into adamantinoma. Osteofibrous dysplasia is typical of children; adamantinoma occurs in adults or adolescents. There are, however, exceptional cases of adamantinoma in children. Osteofibrous dysplasia is almost exclusive of the tibia (and fibula); adamantinoma is prevalent in the tibia but can also rarely occur in other long bones and even in soft tissues. The clinico-imaging presentation of the two lesions can be different, but frequently it is quite similar or identical. Histologically, osteofibrous dysplasia is characterized by a fibro-osseous pattern similar to fibrous dysplasia but containing tiny foci of single epithelioid cells, which can only be seen well using a keratin stain. In an adamantinoma or in an osteofibrous dysplasia transforming into an adamantinoma, small to large nests of epithelioid cells can even be seen on routine H&E staining and can be fully corroborated as epithelioid cells by keratin immunostaining.

Selected Bibliography

See Osteofibrous Dysplasia and Adamantinoma

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