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ADAMANTINOMA OF TIBIA

A CASE REPORT

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SUMMARY:

Adamantinoma is a rare primary malignant tumor of the long bones with unknown pathogenesis. So far only a few cases are reported in the literature (2,3,9,11,17). Tibia is the major site of predilection, however, the tumor also reported occasionally in other long bones such as femur, fibula, humerus, ulna and radius(1,8,12). Most of the patients are in the second and third decades of life. Rarely fibrous dysplasia(5) is associated with adamantinoma of the long bones. The symptoms are long standing. The routine radiologic finding is that of multiple lucent zones interspersed with sclerotic bone,usually with one large rarefied area in the mid shaft.Histologically, islands of epithelial cells with peripheral palisading are seen in fibrous stroma. Histogenesis of

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this tumor is not yet clear. But majority of the studies believe that of epithelial origin (6,15,16,18).Treatment depends upon the extent of tumor. If it is surgically feasible, resection of the tumor is the treatment of choice, if not, amputation. Here we report one case of adamantinoma of tibia and discuss clinicopathological findings.

CASE HISTORY:

A 22-year-old female was admitted in Nemazee Hospital, Shiraz, for the swelling of left leg and difficulty in walking for six months duration. There was a history of minor trauma to the left leg about six years ago. After that, she noticed a small swelling which was growing gradually. But from 6 months prior to admission,the mass started growing rapidly and became painful. All other routine laboratory tests were within normal limits.Chest X-ray was unremarkable.

Clinical examination of the left leg showed a tender cystic mass measuring about 10x8cm. in the anterior aspect of leg. Radiological examination of the left leg showed a large multiloculated cyst in the mid portion of the shaft of tibia with multiple sharply circumscribed lucent zones of different sizes, interspersed by sclerotic bone in between the zones (Fig. 1).

Histological examination of the biopsy material showed multiple small epithelial islands in a fibrous stroma. The epithelial islands varied in size and shape(Fig. II). Frequently they had a peripheral palisding of basal appearing cells with a microcystic center containing loosely arranged cells(Fig.III). A conspicuous feature in so-

me areas was the vascular appearing channels lined by the same kind of cells that formed the epithelial islands (Fig.IV). The epithelial cells appeared relatively inactive with rare mitotic figures. In some fields the fibrous tissue stroma showed cartwheeling appearance.But there is no evidence of fibrous dysplasia in this case. Thus the histologic appearance is diagnostic for adamantinoma of tibia.

DISCUSSION:

Adamantinoma or ameloblastoma is originally described and seen commonly in the jaw bones. It is a true neoplasm, probably arising from enamel organ type tissue. The term adamantinoma indicates that a tumor contains or has the potential of producing enamel. Tumors histologically similar to the ameloblastoma of the jaw are also reported elsewhere in the body; pituitary gland(craniopharyngioma), long bones and soft tissue. But the histogenesis of these tumors is different from that of jaw adamantinoma.

Adamantinoma of long bones is a rare malignant tumor (2,3,9,11,17). The term adamantinoma is misleading for enamel has not been found in these neoplasms. The histologic picture is similar to that of ameloblastoma of the jaw, but the pathogenesis of this peculiar, rare tumor of tibia is the subject of controversy. Whether the trauma plays a part in the histogenesis is not yet clear. One hypothesis given by Changus, Speed and Stewart (4) suggests that adamantinomas have a vascular origin. Another interesting suggestion given by Hicks(7)and by Lederer and Sinclair (10) is that so called adamantinoma of the tibiae is actually synovial sarcoma in disguise.

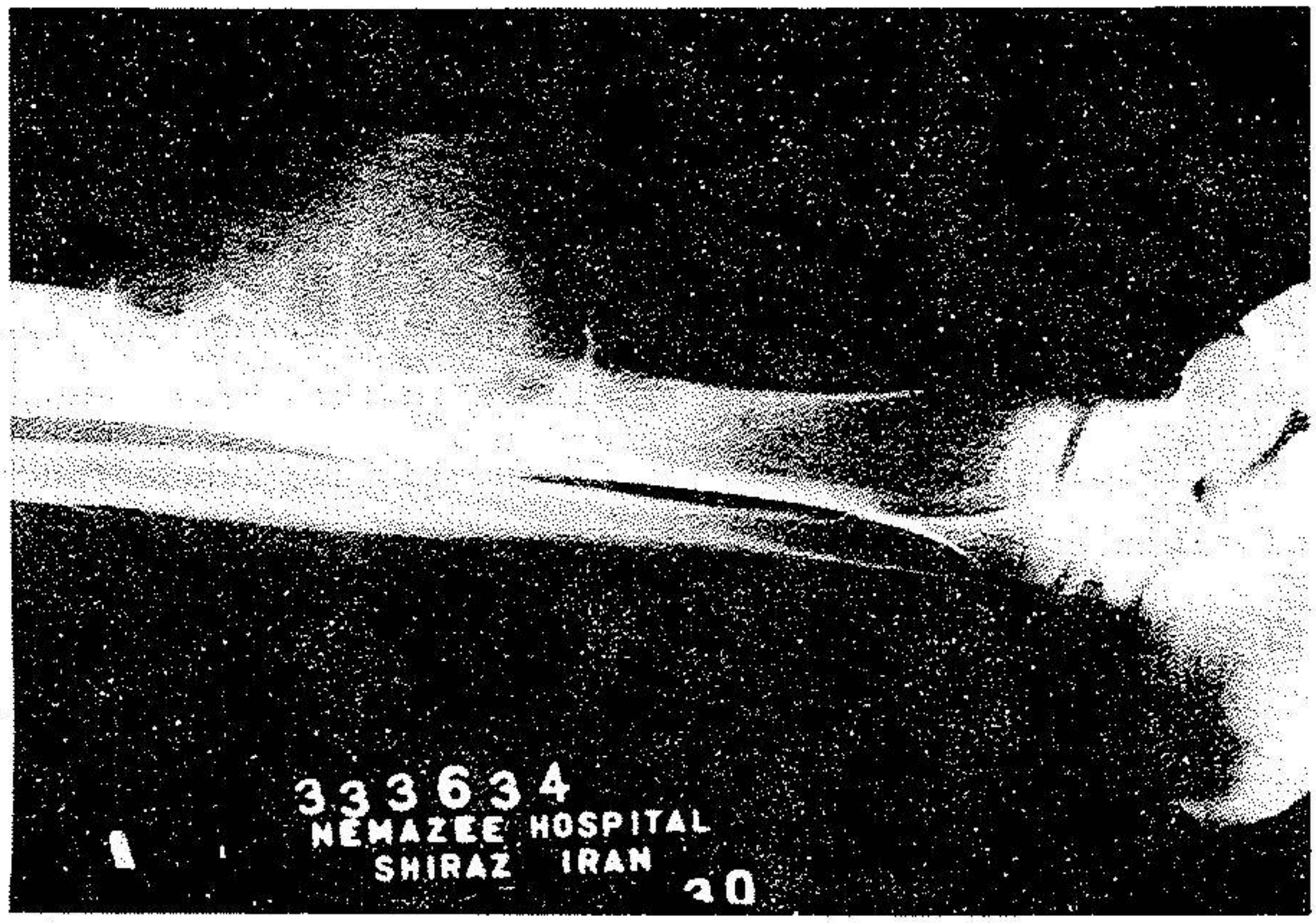

FIG.I: Characteristic radiologic appearance of adamantinoma of the tibia shows multiloculated in the mid shaft of tibia with an admixture of sclerosis.

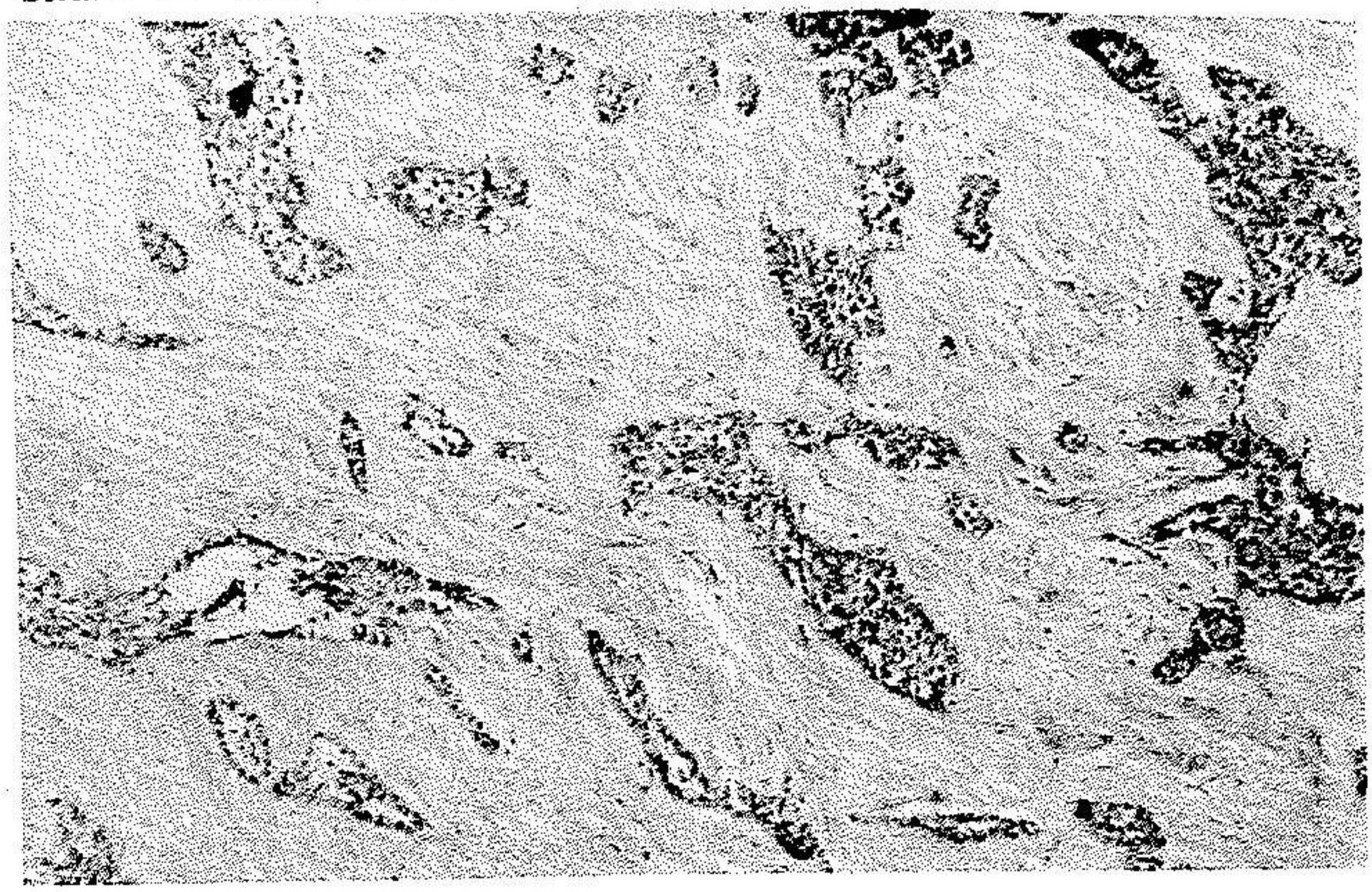

FIG.II: Adamantinoma showing epithelial islands with microcysts in fibrous tissue stroma. H&E:120.

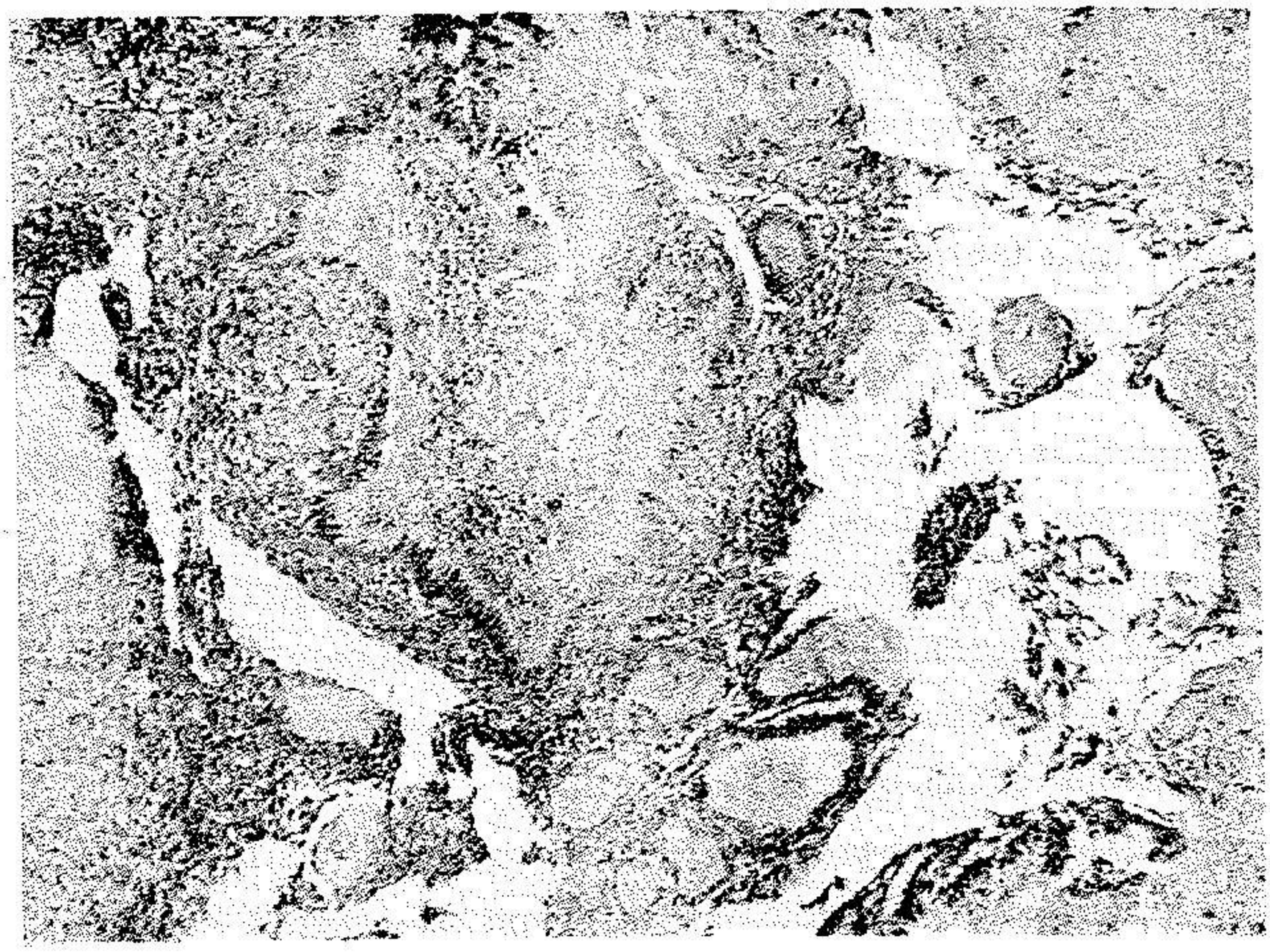

FIG.III: Higher magnification of epithelial islands show peripheral palisading of cells. H&E:450.

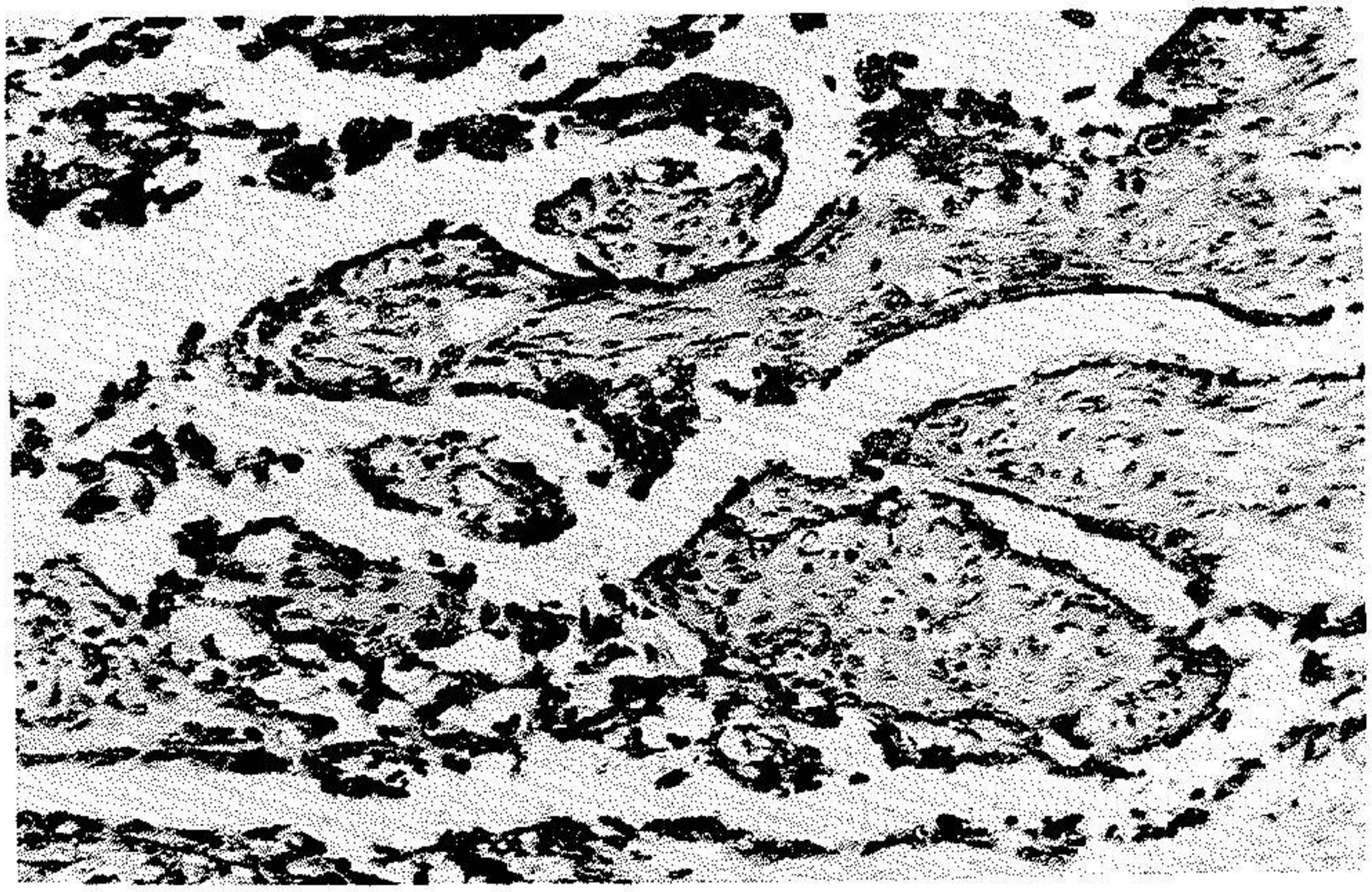

FIG.IV: Adamantinoma showing vascular like channels lined by the same kind of cells that formed epithelial islands. H&E: 450.

Cohen (5) theory suggests that these tumors have their origin in misplaced embryonal rests (dermal inclusion tumors). Most of the electron microscopic studies of tibial adamantinoma (15,17, 18) have supported the concept of an epithelial origin, except, Povysil and Matejovsky (14) studies which are in favour of mesodermal origin. Immunohistochemical studies (6,16) showed positive carcinoembryonic antigen, keratin and negative factor VIII related antigen, which support an epithelial origin. The recent electron microscopic study (6) disclosed desmosomes and intermediate sized filaments, which are strongly in favour of an epithelial origin.

Clinically these tumors develop insidiously and slowly often over a period of years and manifest a tendency to progressive local onvasion.Some cases have shown that the tumors are capable of extension to regional lymph nodes and even metastasis to the lungs(13). The tumors might produce pain, swelling and occasionally pathologic fracture. Radiographically the tumor appears as a smaller or larger, finely trabeculated, cystic well defined mass without cortical destruction. The tumor may attain a large size.

Histologic picture may vary from specimen to specimen and even in different fields of the same specimen.In some places one may observe glandular pattern which may be mistaken for metastatic adenocarcinoma and causes diagnostic problem(17). In such situation, examination of additional material is useful. In other places, the tumor may be resembled to basal cell carcinoma, growing in strands and (or) nests. In some recorded cases, the histologic picture showed frank pearl formation,so that it may be mistaken

for squamous cell carcinoma (18). The classical histologic picture of adamantinoma is the presence of epithelial cell islands in a fibrous tissue stroma. The epithelial islands show microcystic degeneration with peripheral palisading basal appearing cells. Thus the histologic picture of this tumor is reminiscent of ameloblastoma of the jaw.

Treatment of this rare tumor is again a matter of controversy. A group of people have suggested amputation, others have advocated resection (2,11), Unni et al(17) believe that when technically feasible, resection of the lesion is the treatment of choice otherwise amputation. The role of radiotherapy is not clear in the treatment of this tumor, however, apparent cure of the tumor with radiotherapy has been reported (19).

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