Atypical "Sclerosing" Osteoblastic Neoplasm

A Tumor of Intermediate Biological Potential Between Usual Osteoblastoma and Conventional Osteosarcoma

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Osteogenic tumours

Benign

Osteoma

Osteoid osteoma

Intermediate (locally aggressive)

Osteoblastoma

Malignant

Low-grade central osteosarcoma

Conventional osteosarcoma

- Chondroblastic osteosarcoma
- Fibroblastic osteosarcoma
- Osteoblastic osteosarcoma

Telangiectatic osteosarcoma

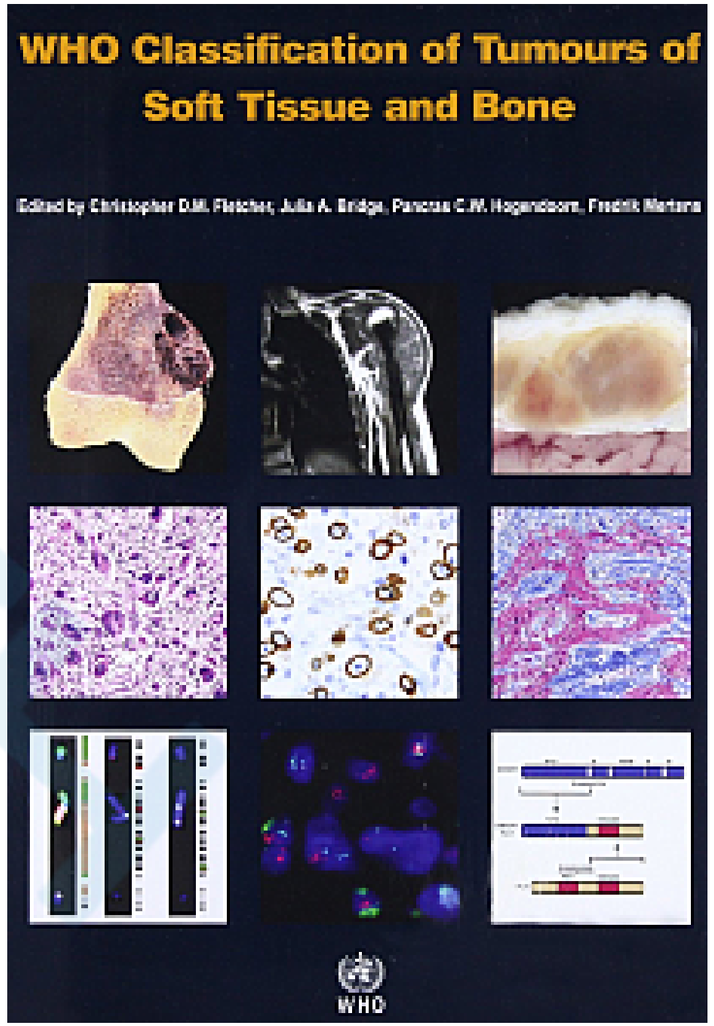
Small cell osteosarcoma

Secondary osteosarcoma

Parosteal osteosarcoma

Periosteal osteosarcoma

High-grade surface osteosarcoma



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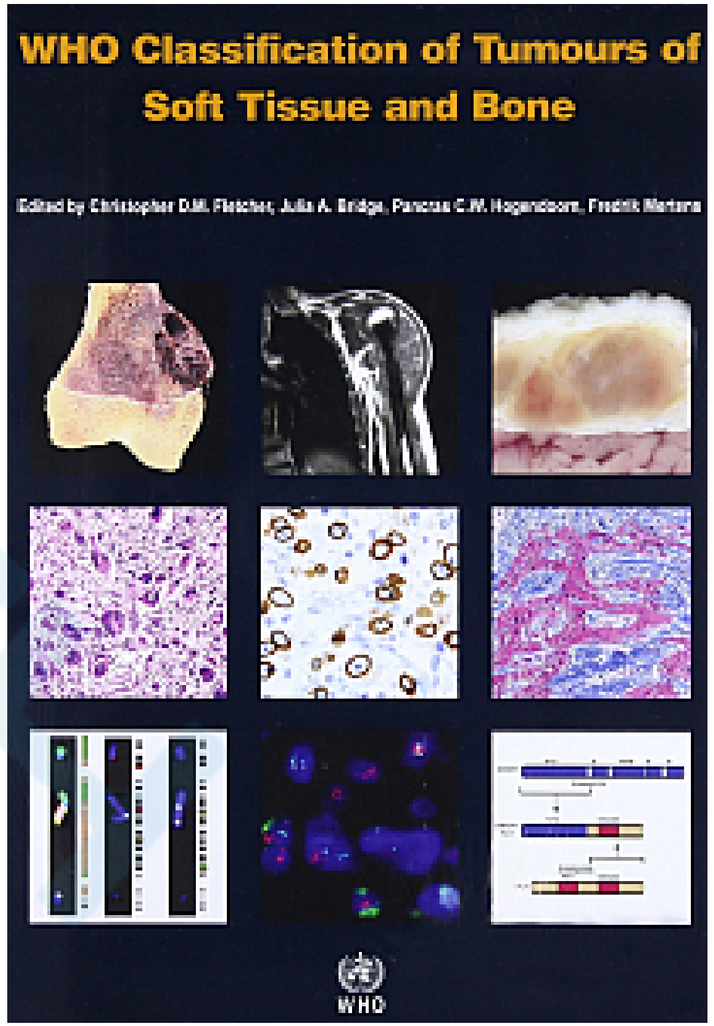

Osteoblastoma（OB）

Definition

A benign bone-forming neoplasm, > 2 cm, which produces woven bone spicules, which are bordered by prominent osteoblasts

- Incidence: 1%
- Gender: M : F = 2.5 : 1
- Age: 10～30（5～70）
- Site:

Posterior elements of spine and sacrum (40～55%)

Proximal femur, distal femur and proximal tibia

Background

- Conventional OB is considered a benign neoplasm, lacking the ability to metastasize, though approximately one fifth of tumors do locally recur
- Whether OB can transform or dedifferentiate into an osteosarcoma (OS) acquiring metastatic potential, remains controversial
- The recognized histomorphologic overlap of OB with OS, particularly the relatively recently described OB-like OS, adds further to this controversy

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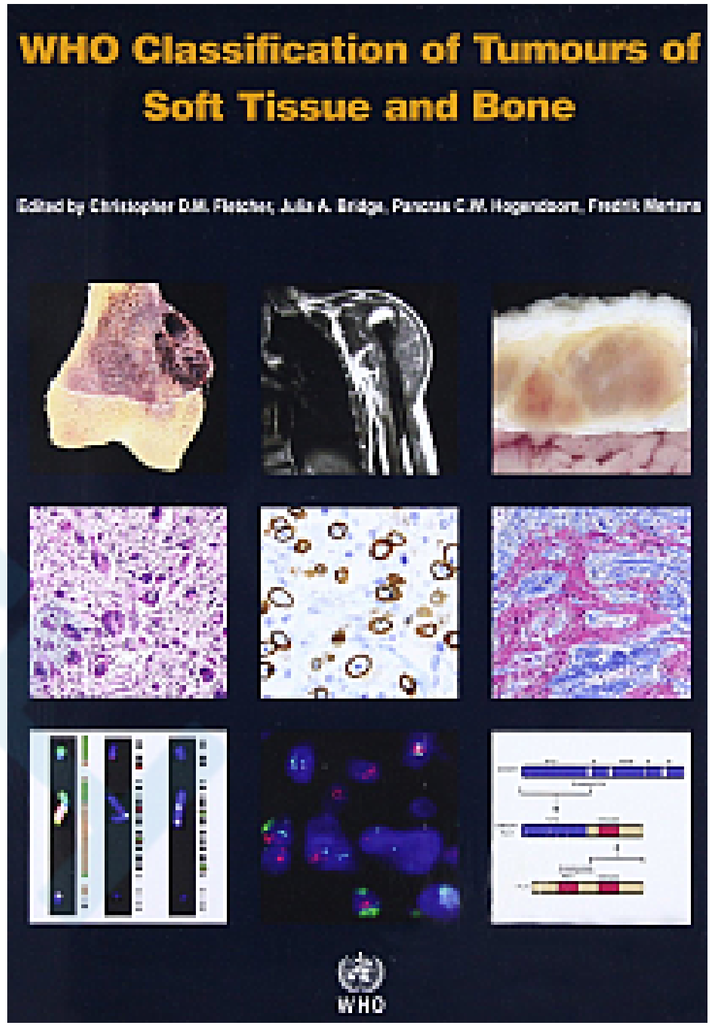


Table 16.01 Histological subtypes of osteosarcoma

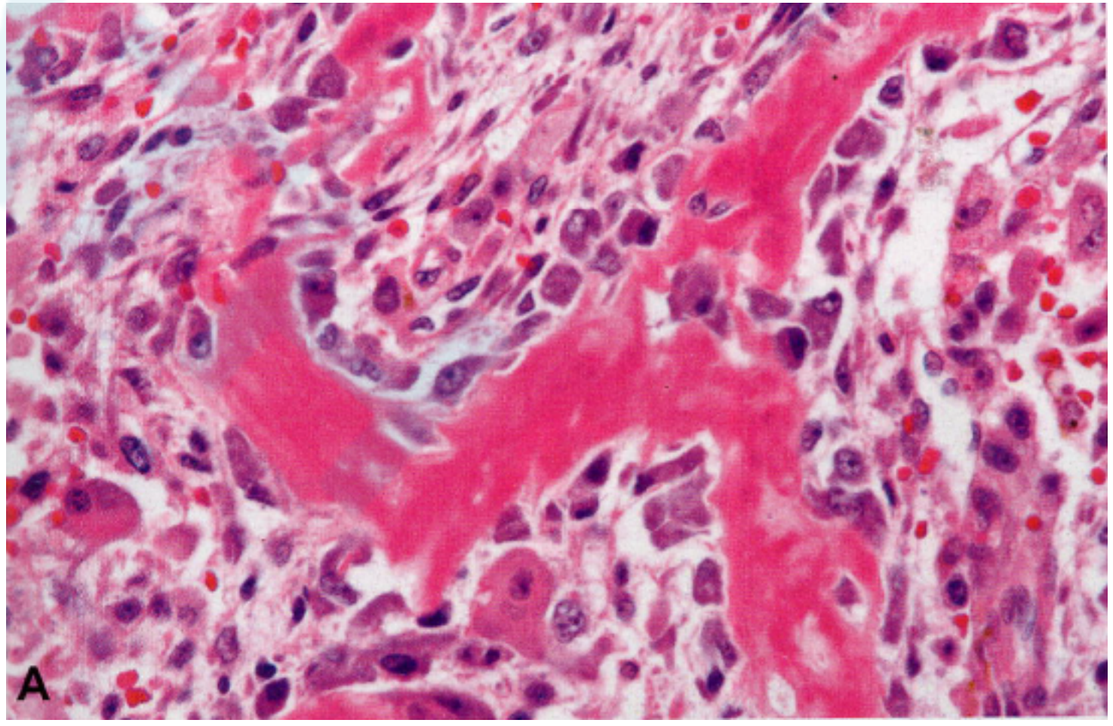
Osteoblastic (including sclerosing)
Chondroblastic
Fibroblastic
Giant cell rich
Osteoblastoma-like
Epithelioid
Clear cell
Chondroblastoma-like

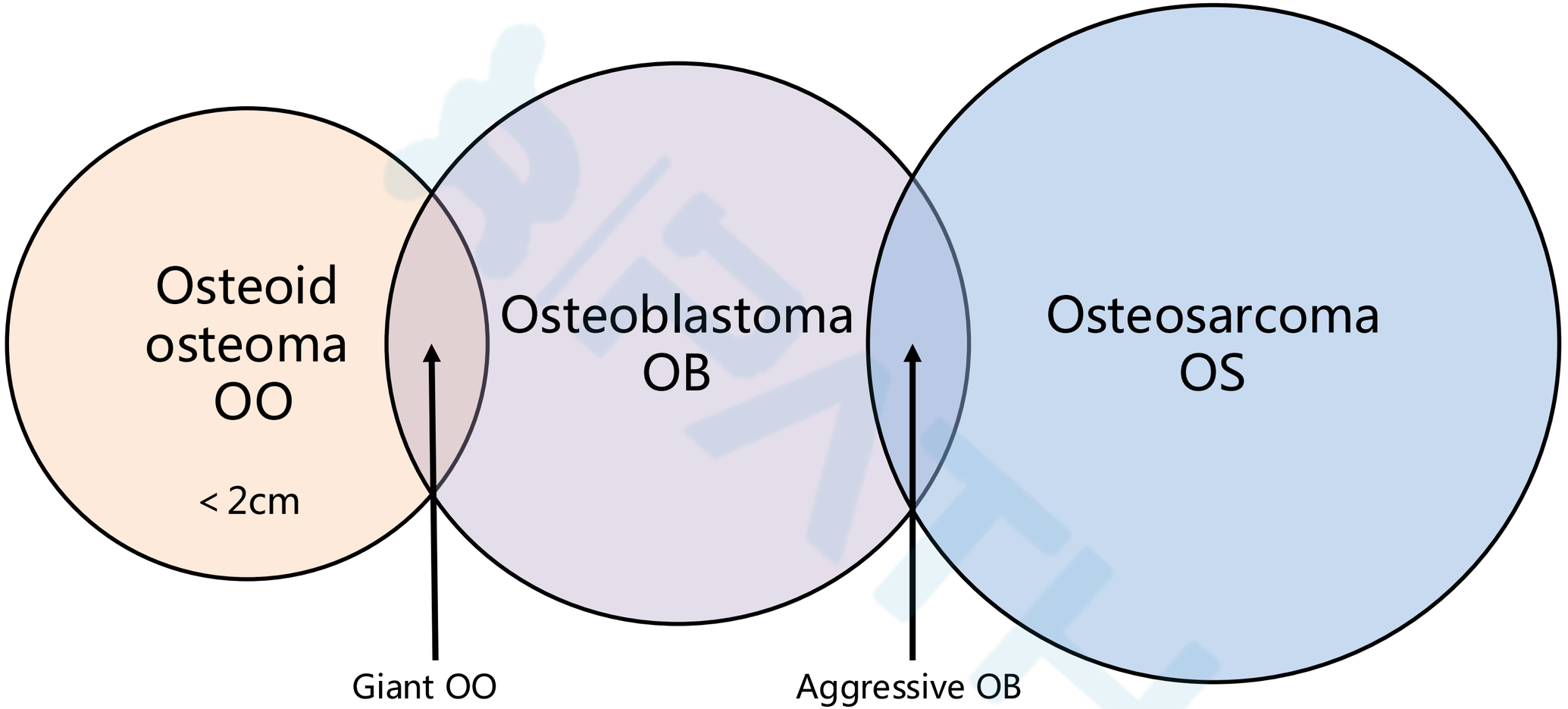
OB-like OS

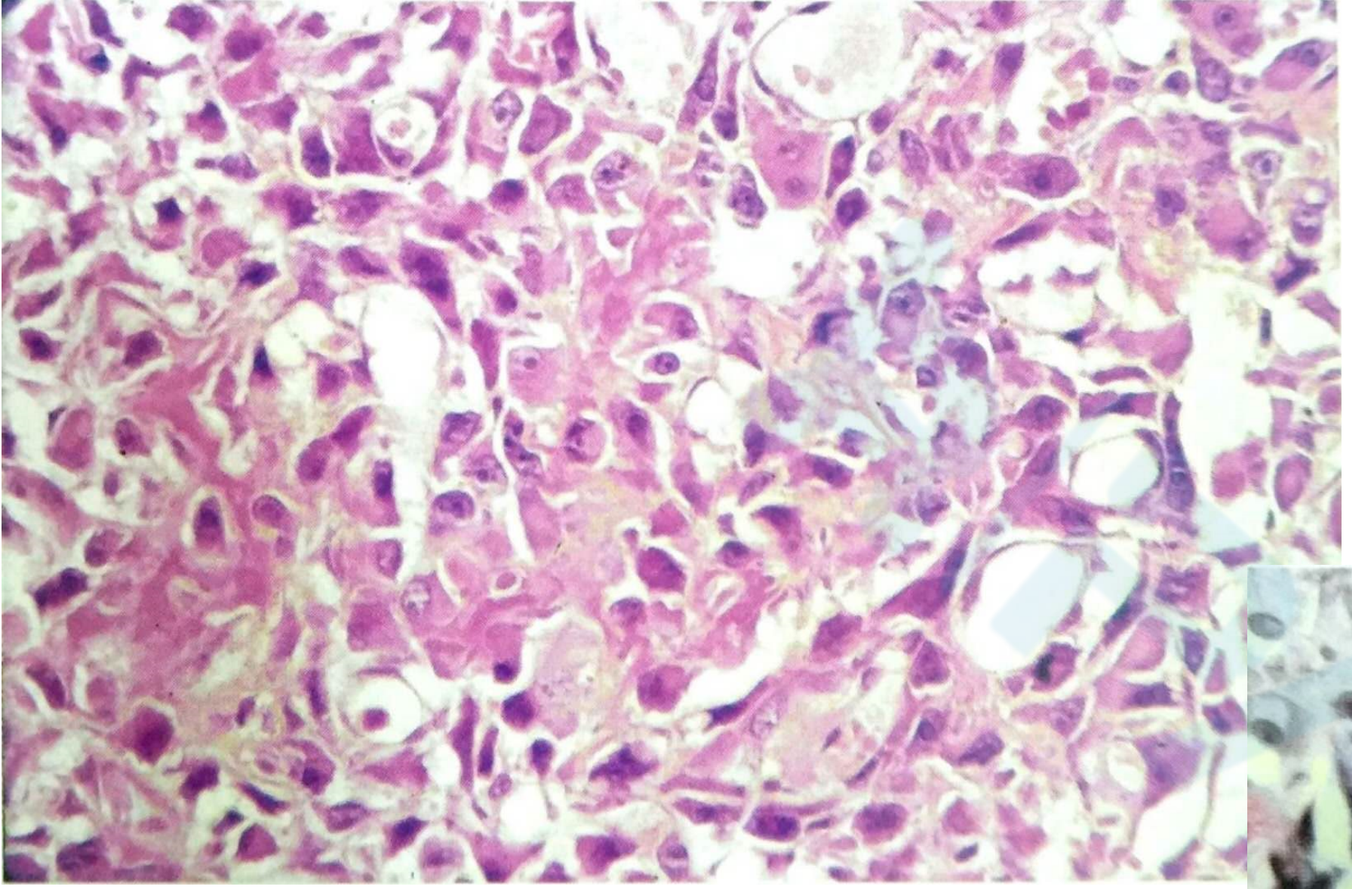
acterize the epithelioid variant {1474}. In the osteoblastoma-like variant, the tumour cells may rim the neoplastic bony trabeculae in a fashion that mimics osteoblastoma. Features that permit its distinction are the permeative growth pattern, cells that are cytologically atypical and cellular intertrabecular regions {204}.The chondro-

OB-like OS is a rare variant of OS

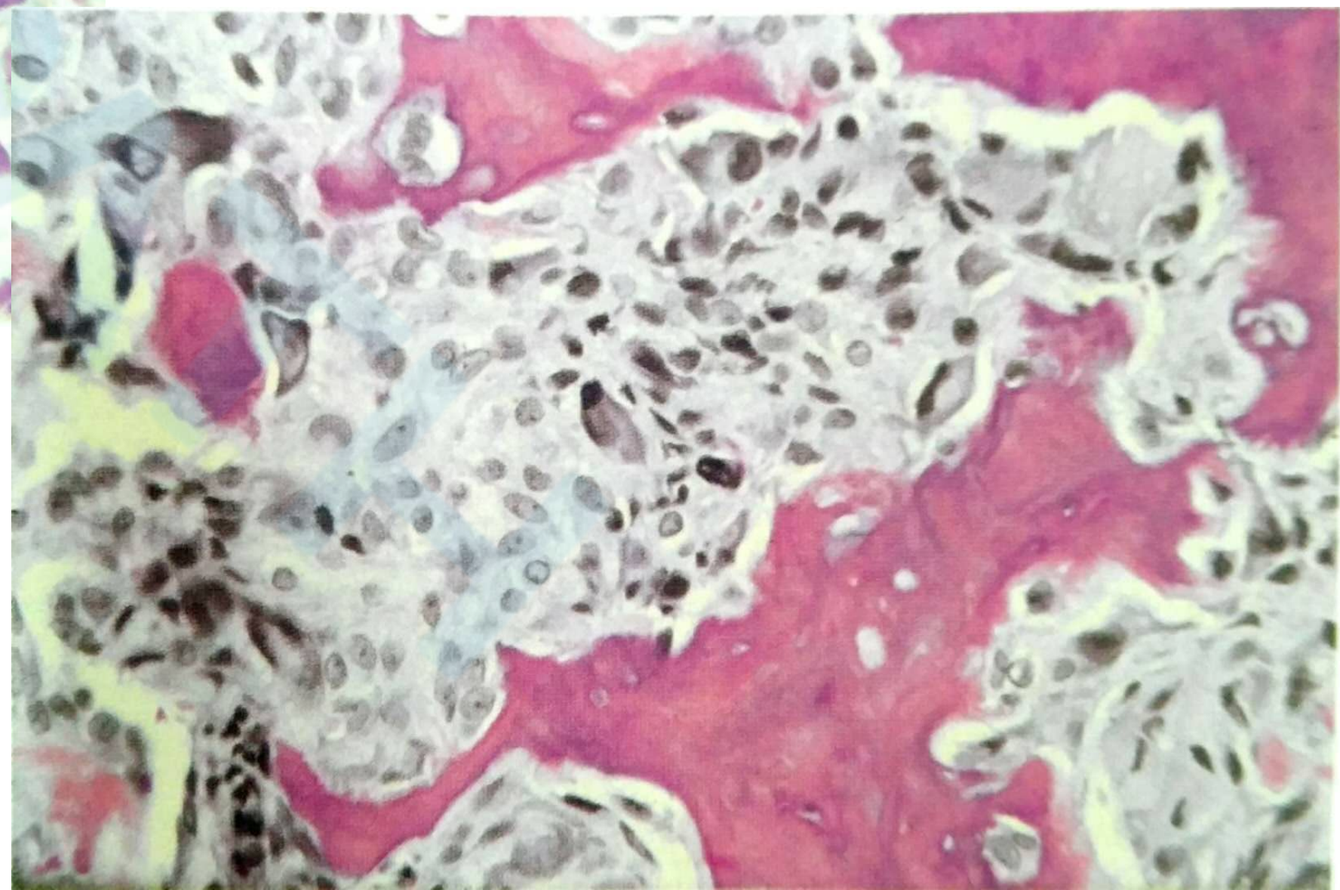
- Sometimes described as a high-grade malignancy and at other times as a low-grade neoplasm, largely based on differing clinical behavior
- The pathologic criteria that define it include peripheral permeation of the neoplasm into the surrounding bone, cellular sheets of tumor cells devoid of vascular stroma, and an elevated mitotic rate (3-10/10HPF)



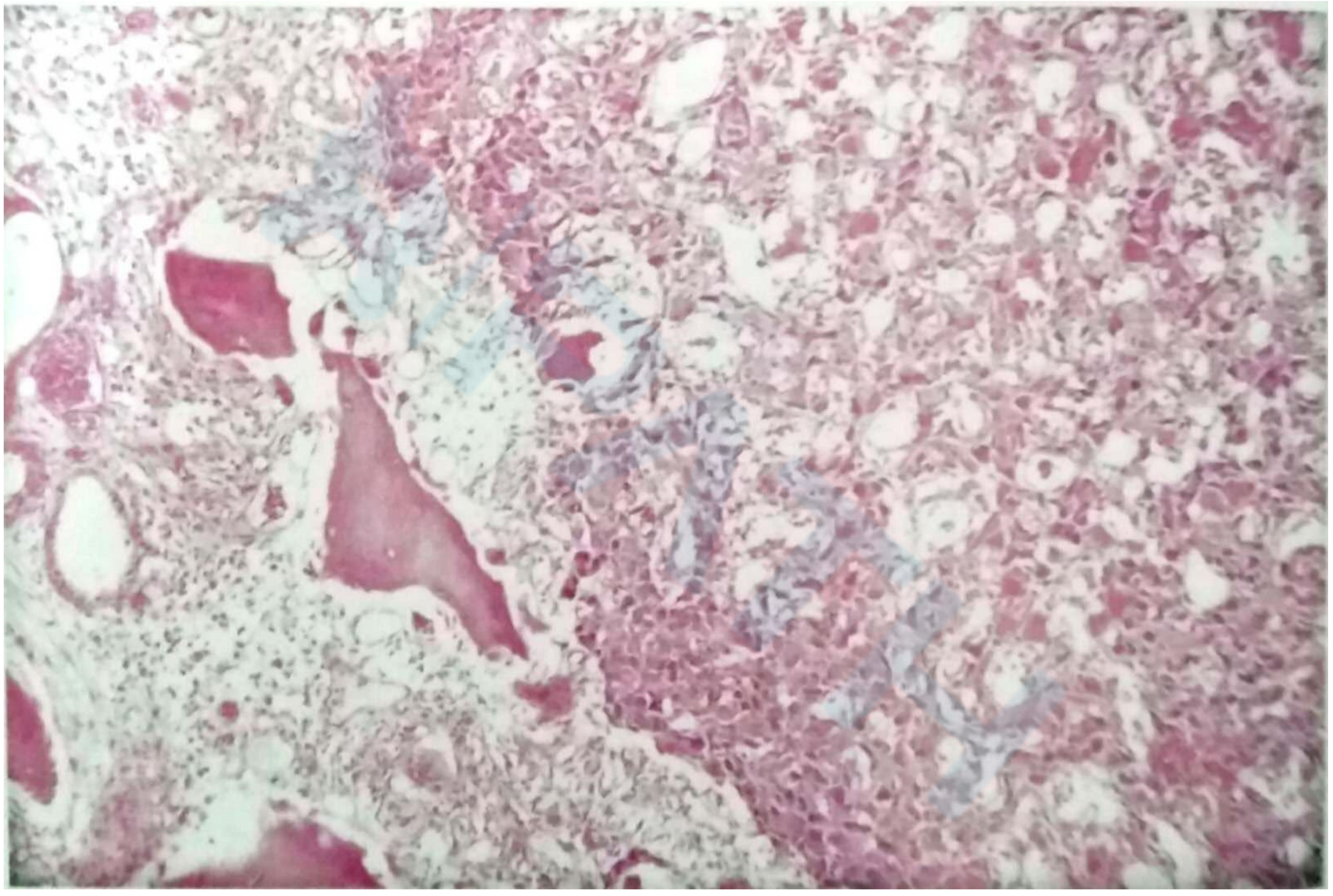

Osteoid
osteoma
OO
< 2cm
Osteoblastoma
OB
Osteosarcoma
OS
Giant OO
Aggressive OB



OB-like OS



Aggressive OB



A group of bone-forming tumors

Histologically

- Distinct from conventional OB
- Not typical of aggressive OB
- Not meeting the criteria for OS

Microscopically

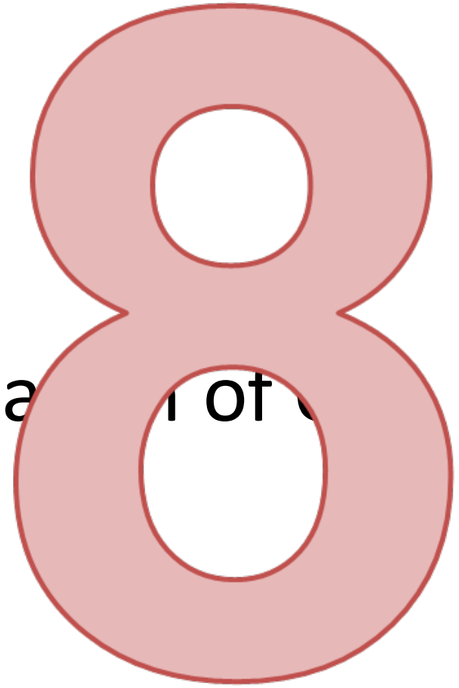
- Compact, sclerosing sheet-like neoplastic bone
- Few osteoblasts
- Minor OB-like areas

Radiographic

- Not helpful in predicting behavior
- Typically lacked aggressive features

MATERIAL AND METHODS

- Over 22 years
- 3 co-authors
- Diagnoses of atypical OB, malignant transforma n of
  well-differentiated OS, and OB-like OS
- Tumors had to demonstrate



Majority: predominantly compact, sclerosing sheet-like pattern of neoplastic bone with few osteoblasts

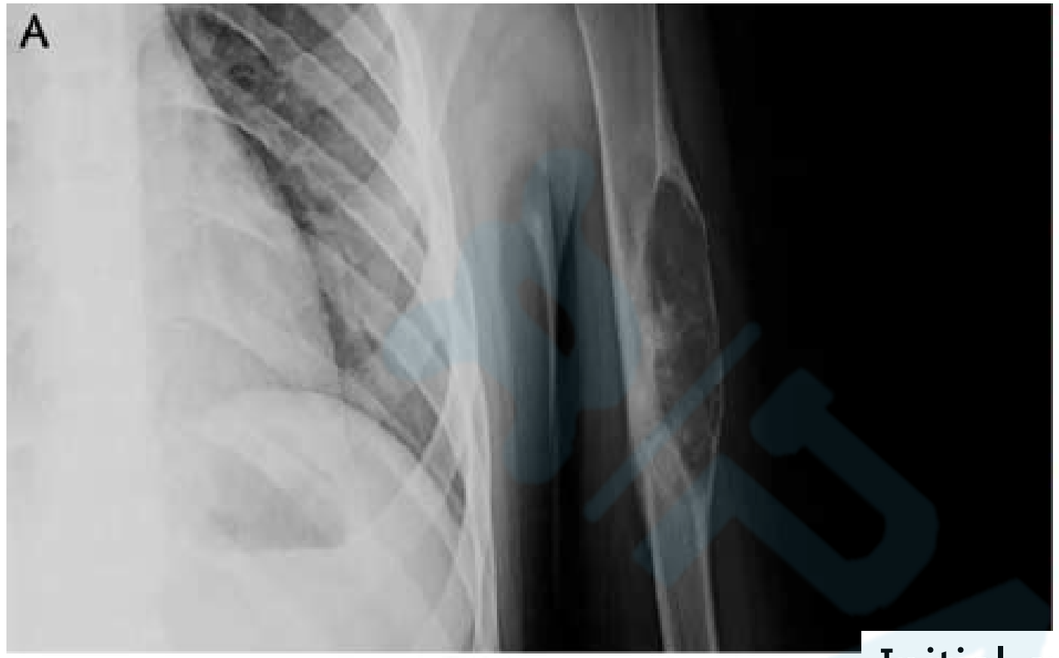
Minor: conventional OB like areas, without any evidence of permeation

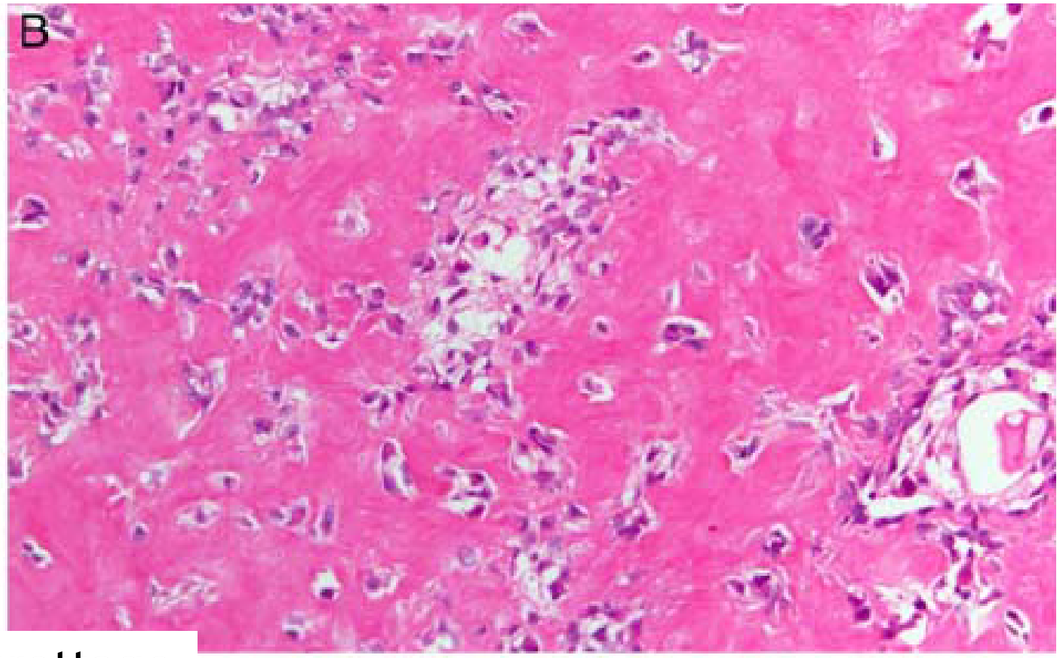
RESULTS

TABLE 1. Clinical and Histopathologic Features of Atypical Sclerosing Osteoblastic Neoplasms

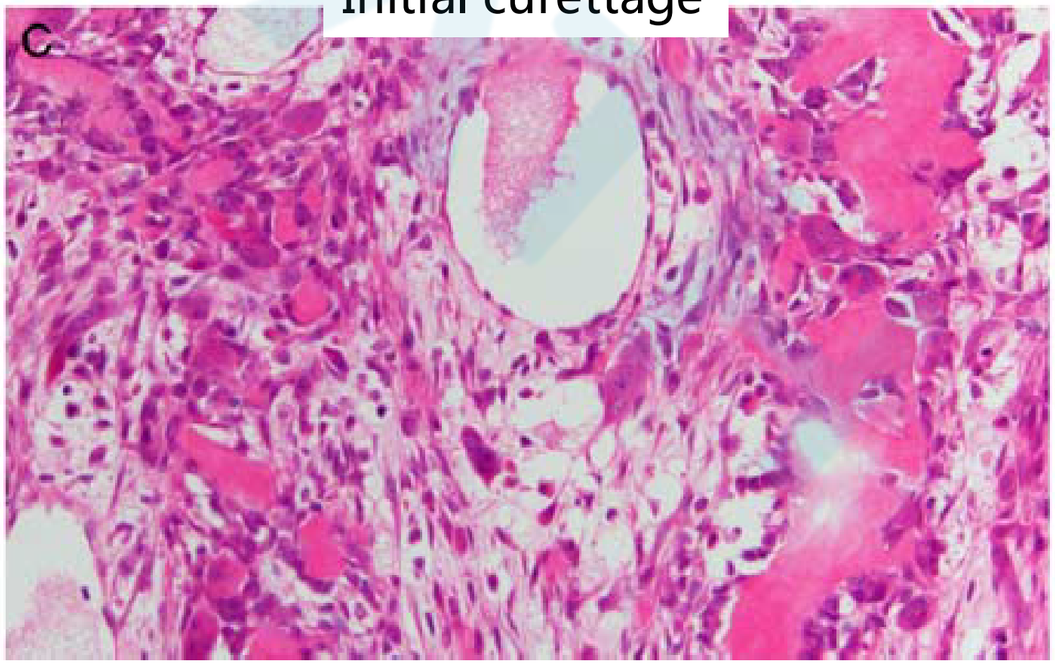
Case	Age (y)/ Sex	Site	Initial Diagnosis	Initial Treatment	Follow-up
1	12/M	Left humerus	Osteoblastoma*	Curettage	Local recurrence, 8 mo. Dx rendered "atypical osteoblastic lesion, most consistent with osteosarcoma." Lost to follow-up, 1 y
2	11/M	Left femur	Unusual bone-forming lesion, most consistent with osteosarcoma*	En bloc intralesional resection	Local recurrence, 2 y. Dx rendered "atypical osteoblastic lesion, most consistent with osteoblastoma-like osteosarcoma." AWD, 3 y 10 mo
3	22/M	Right fibula	Osteoblastoma	Curettage	Local recurrence, 4 mo. Dx unchanged. En bloc resection with negative margins. NED, 1.5 y
4	25/F	Right fibula	Subperiosteal sclerosing osteoblastoma*	Curettage	Local recurrence, 3 y 1 mo. Dx unchanged. En block resection, osteoblastoma* with negative margins. NED, 11.5 y
5	38/F	Right 2nd metatarsal	Osteoblastic osteosarcoma*	Curettage & en bloc resection, negative margins	Considered a low grade osteosarcoma. NED, 9.5 y
6	55/M	Left 3rd metatarsal	Osteoblastoma	Curettage & en bloc resection, negative margins	Local recurrence, 3 y. Dx rendered "osteosarcoma" with comment that it may represent "malignant degeneration of an osteoblastoma." BKA, lost to follow-up. DOD, 11 y
7	27/M	Tibia	Osteosarcoma*	BKA	BKA performed after initial diagnostic biopsy. NED, 3 y
8	16/M	Right 1st metatarsal	Atypical sclerosing osteoblastic neoplasm*	Curettage	No local recurrence as of 9 mo. Patient actively being followed

Case 1



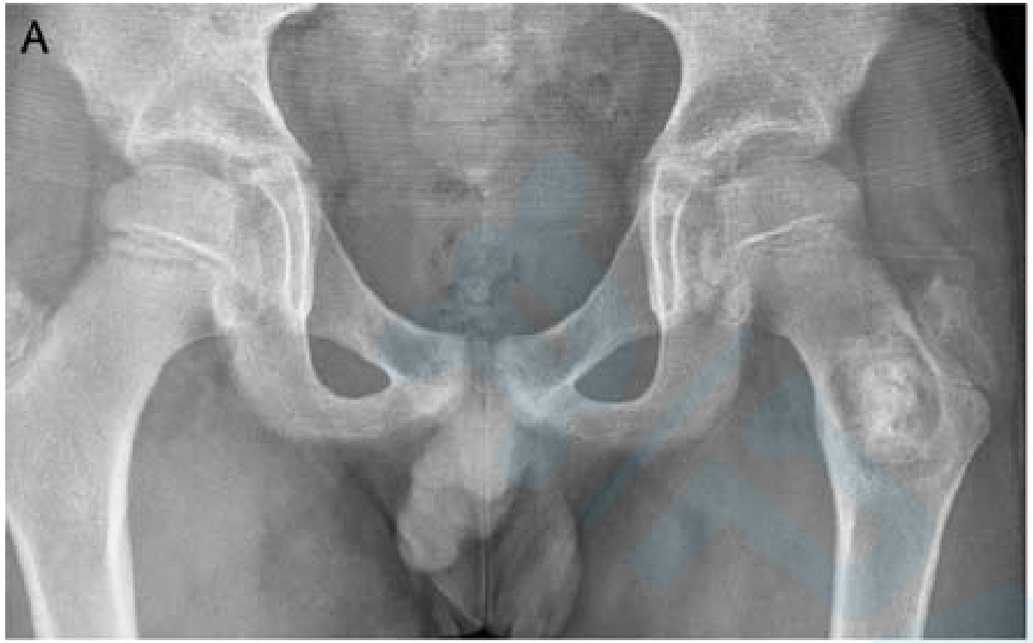


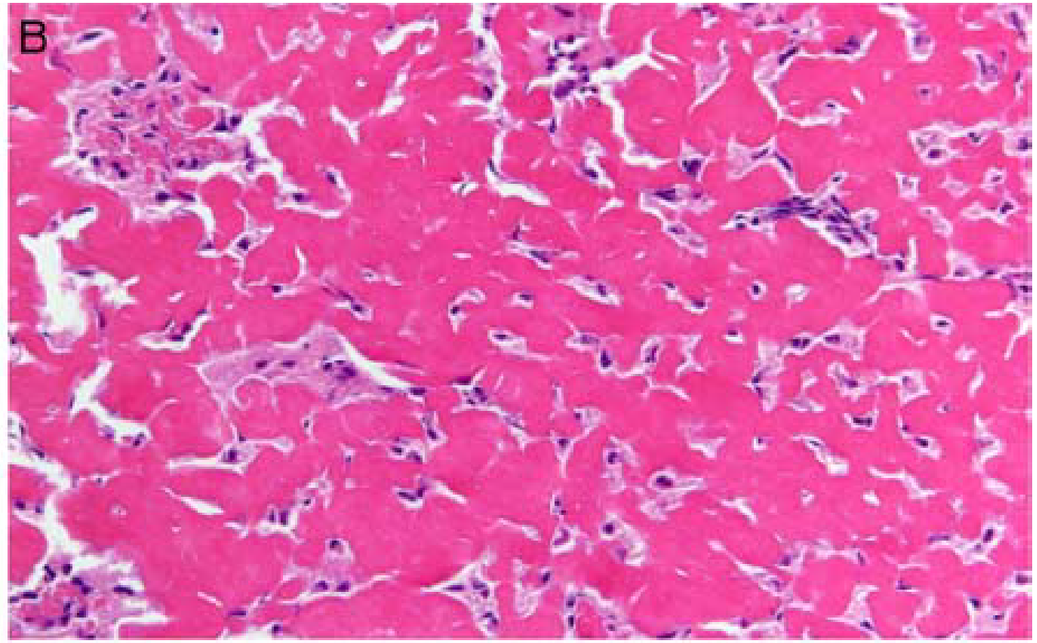
Initial curettage



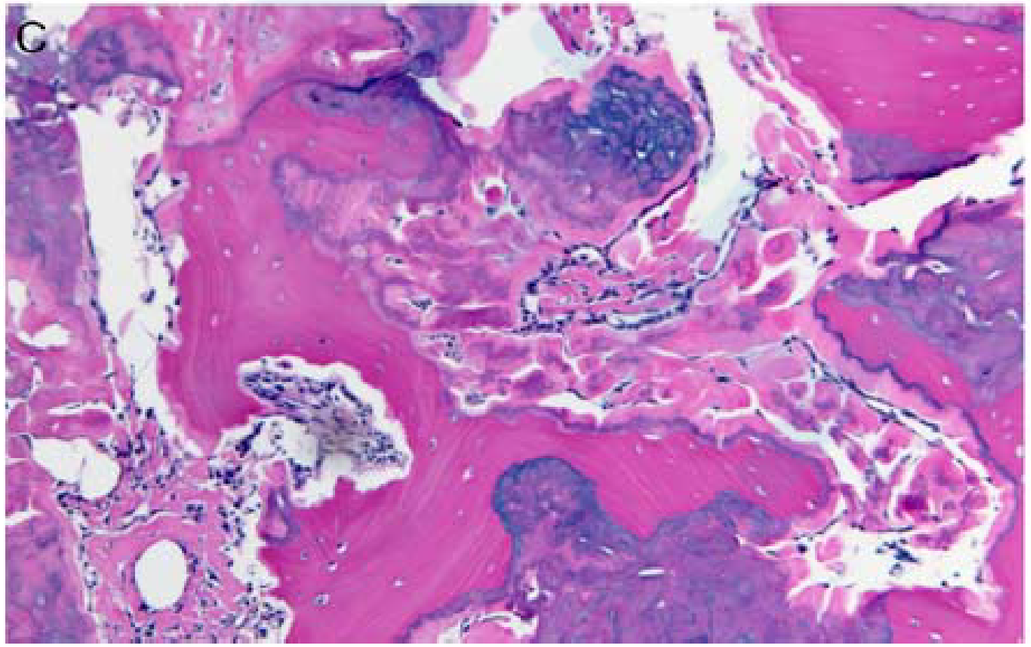
Local recurrence, 8 mo.
Dx rendered “atypical osteoblastic lesion, most consistent with osteosarcoma.”
Lost to follow-up, 1 y

Case 2





Initial curettage
"unusual bone-forming lesion"

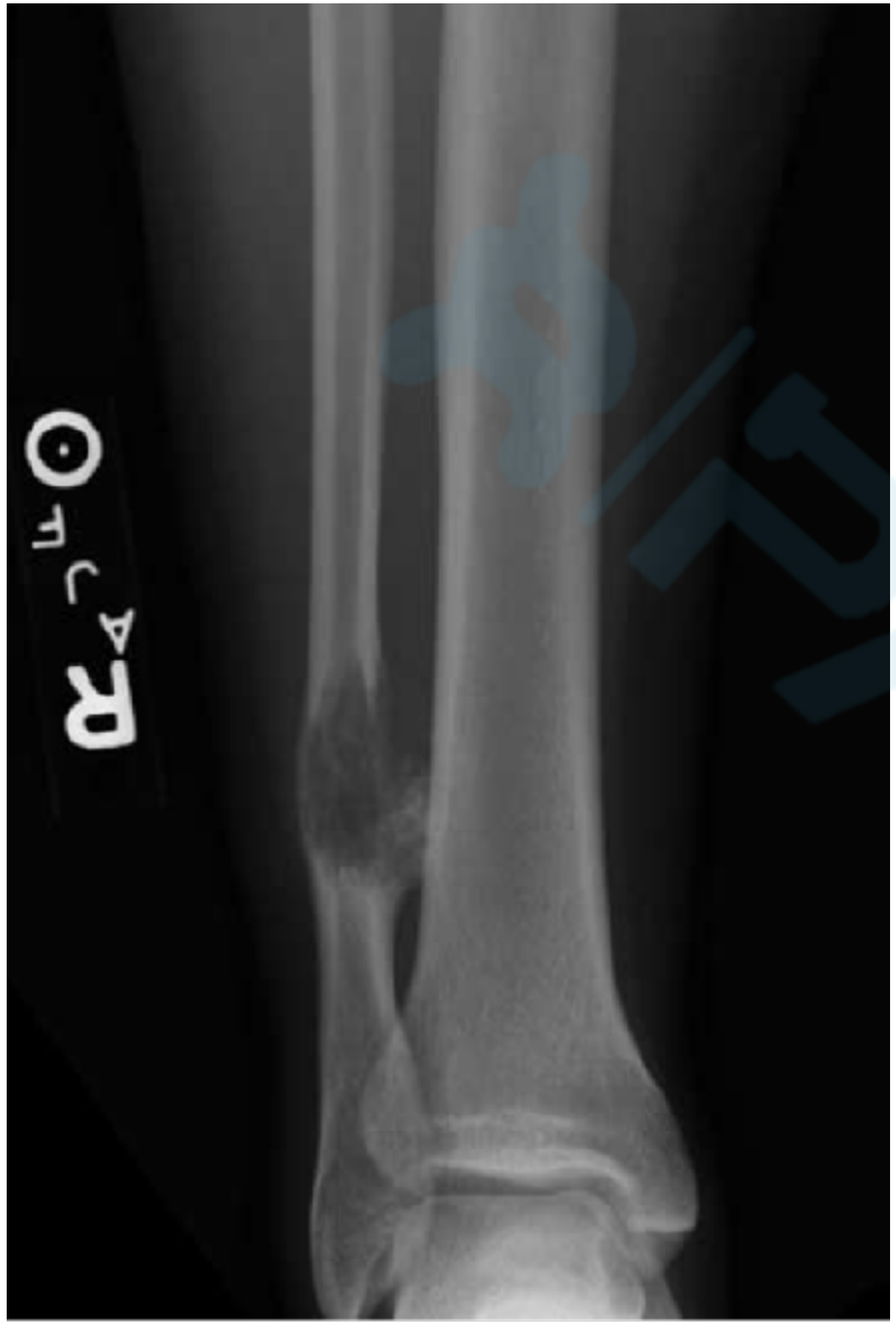


Local recurrence, 2 y
AWD*, 3 y 10 mo

Local recurrence
En bloc intralesional resection
Focally permeative pattern
"Atypical osteoblastic lesion, most consistent with osteoblastoma-like osteosarcoma"

*AWD indicates alive with disease

Case 3

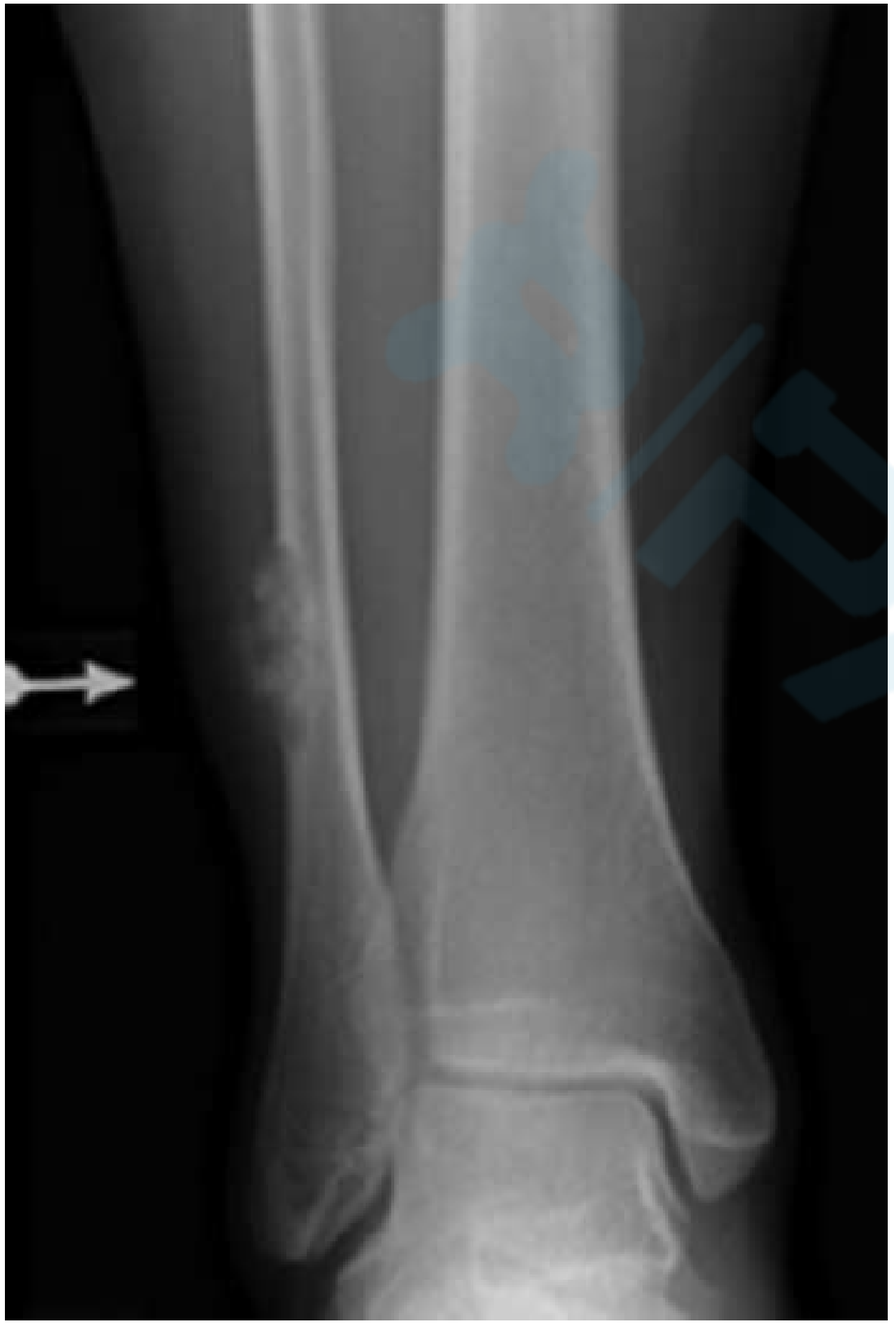


Curettage
Local recurrence, 4 mo. Dx unchanged
En bloc resection with negative margins
NED*, 1.5 y

*NED, no evidence of disease

“osteoblastoma” with soft tissue extension

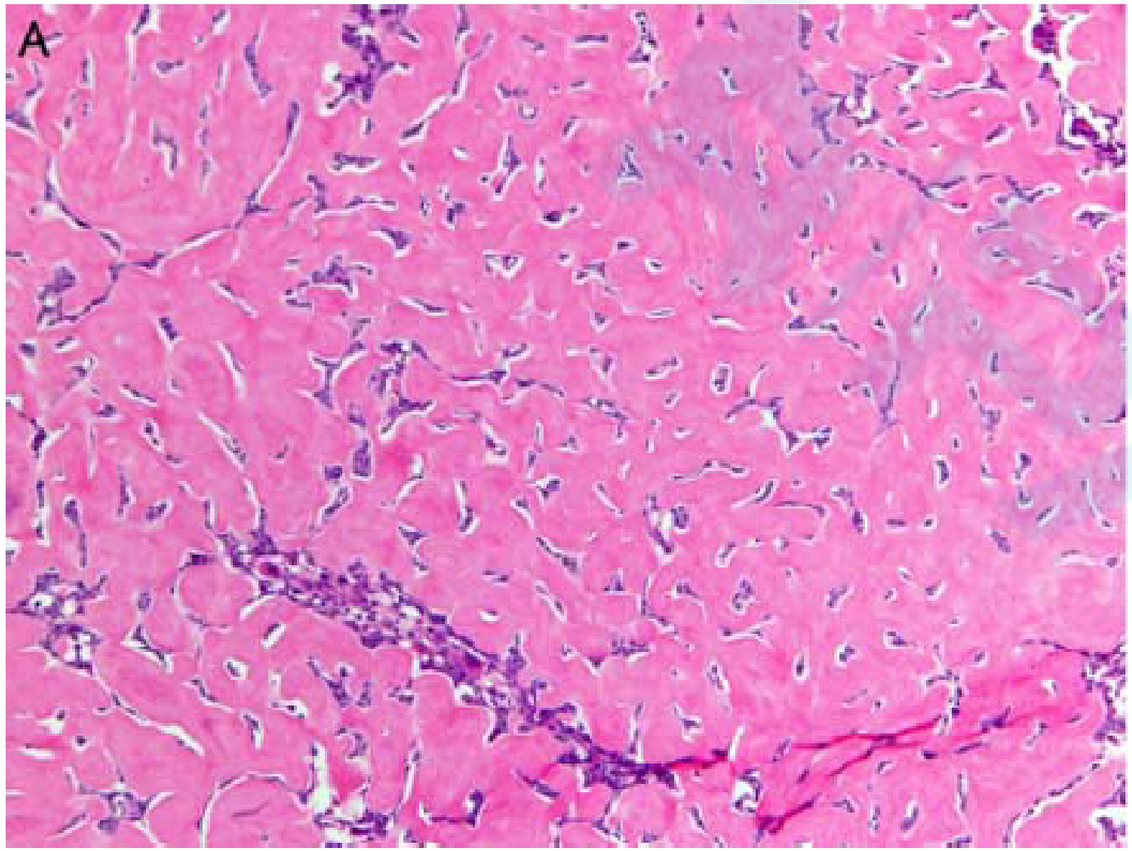
Case 4

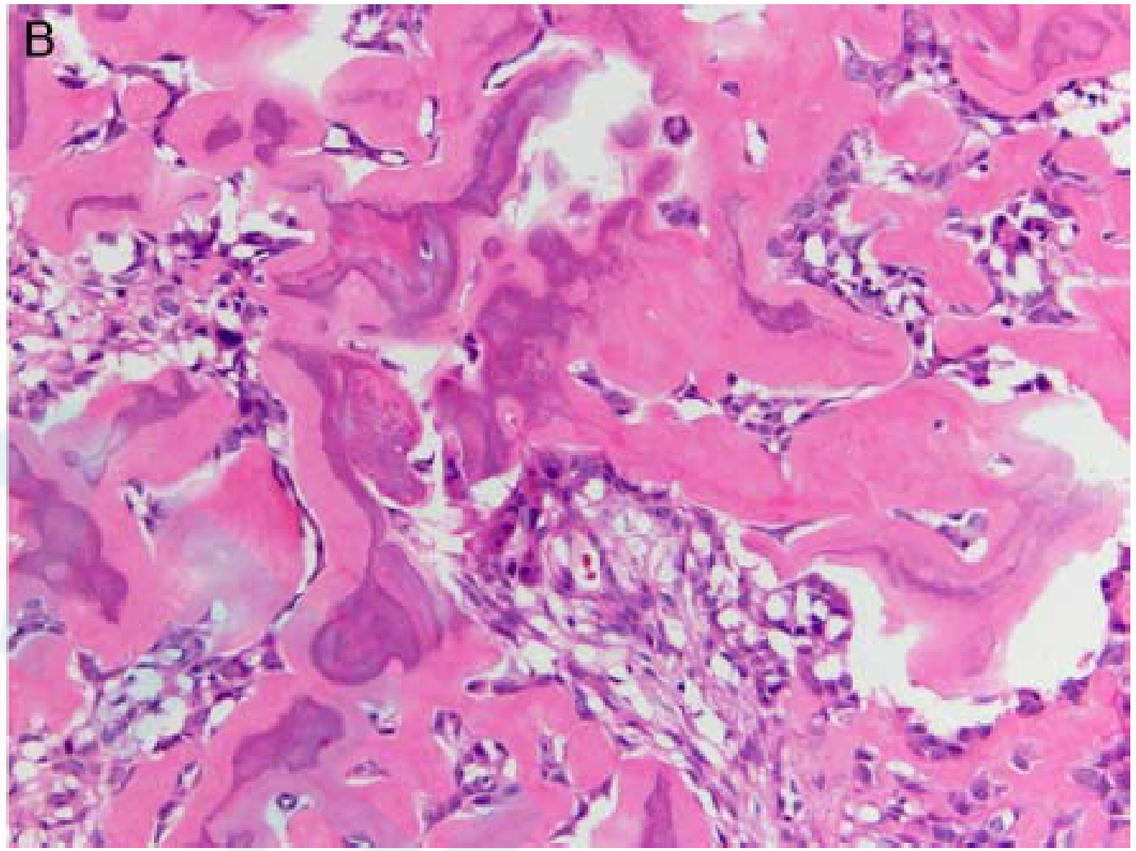


Curettage
Local recurrence, 3 y 1 mo. Dx unchanged
En block resection with negative margins
NED, 11.5 y

"Subperiosteal sclerosing osteoblastoma" appears circumscribed and nonaggressive

Case 5



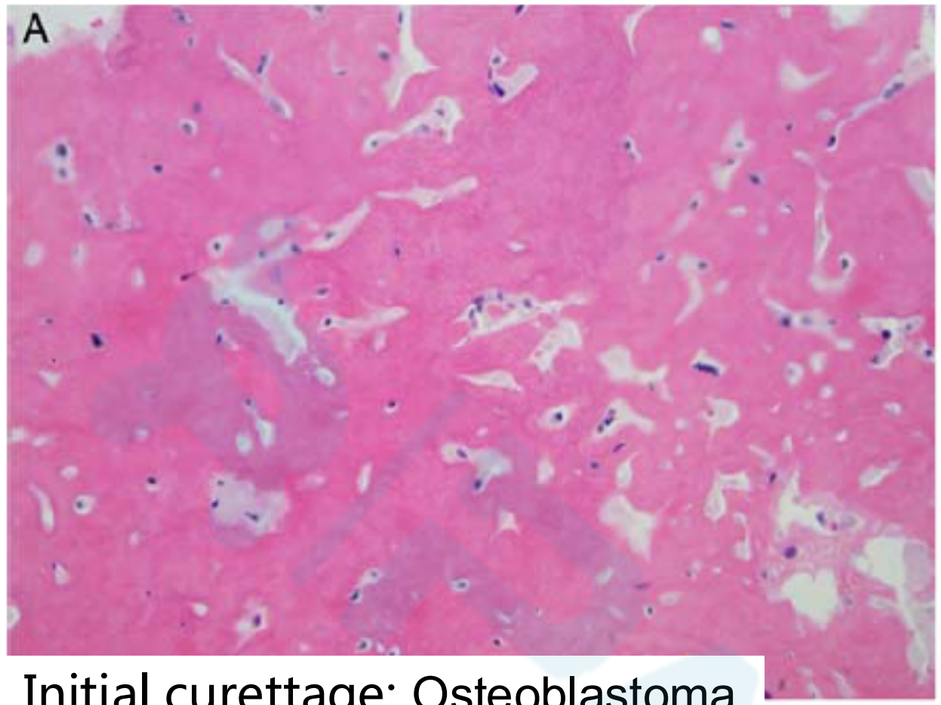


Initial Diagnosis: “osteoblastic osteosarcoma”

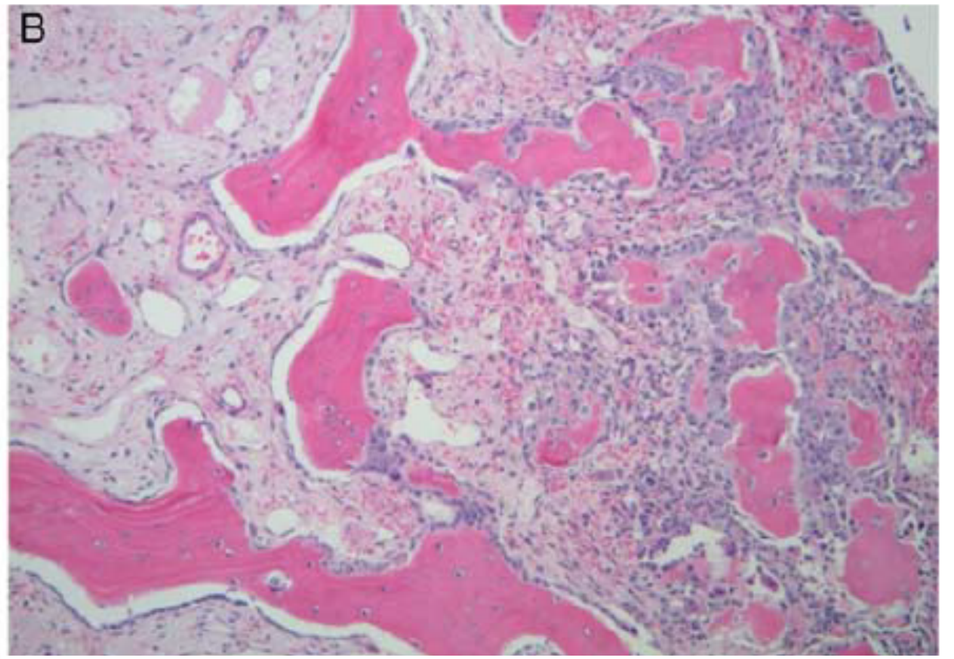
A minority of foci were more typical of osteoblastoma

Curettage & en bloc resection, negative margins
Considered a low grade osteosarcoma. NED, 9.5 y

Case 6



Initial curettage: Osteoblastoma

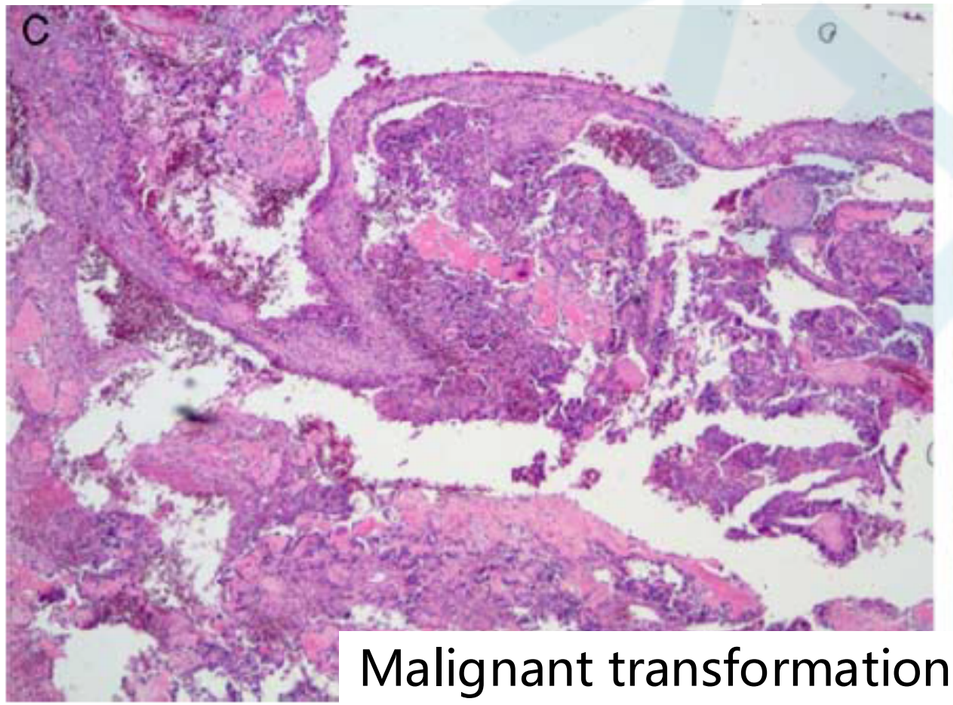


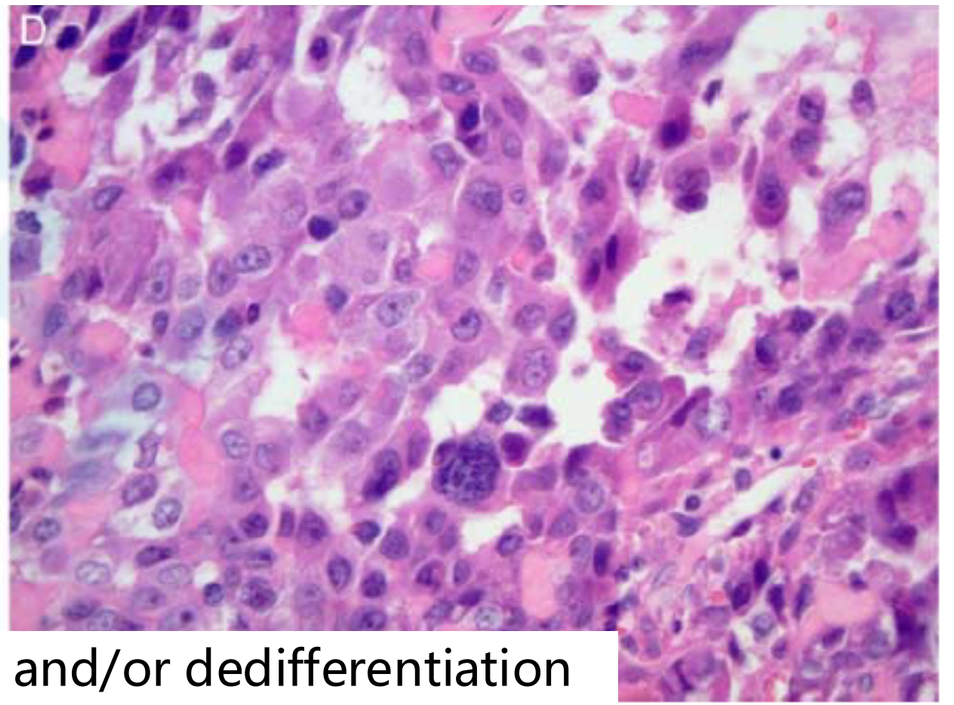
Classic osteoblastoma-type appearance

Curettage & en bloc resection, negative margins

Local recurrence, 3 y
BKA*, lost to follow-up
DOD*, 11 y

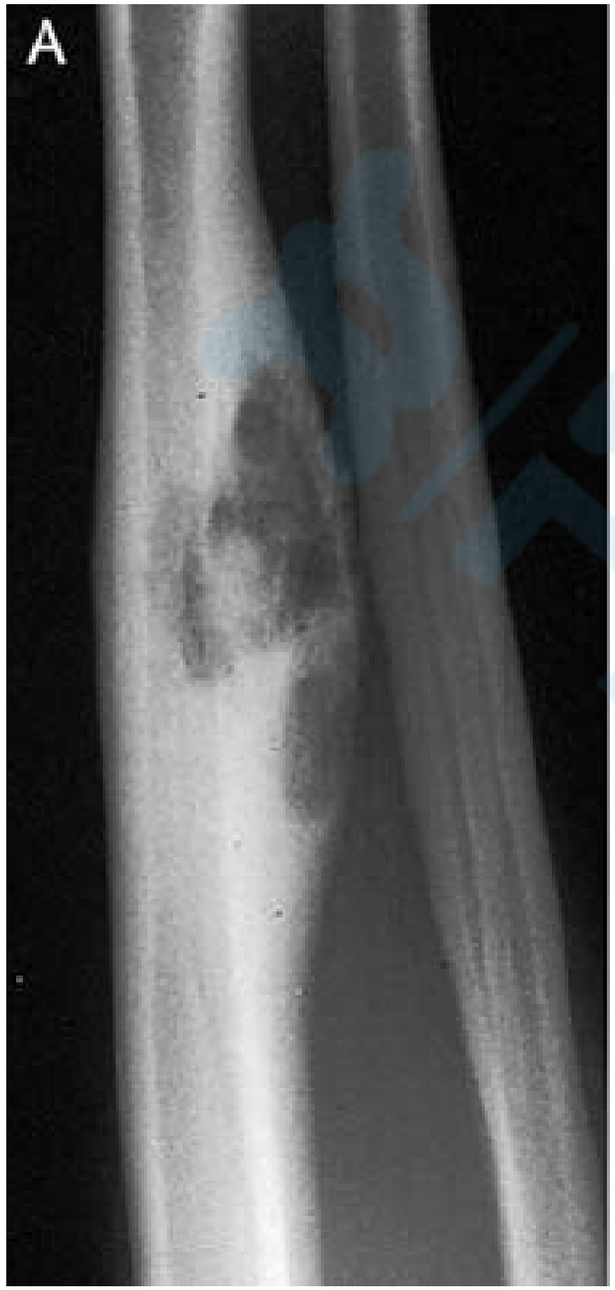
*BKA, below knee amputation
*DOD, dead of disease

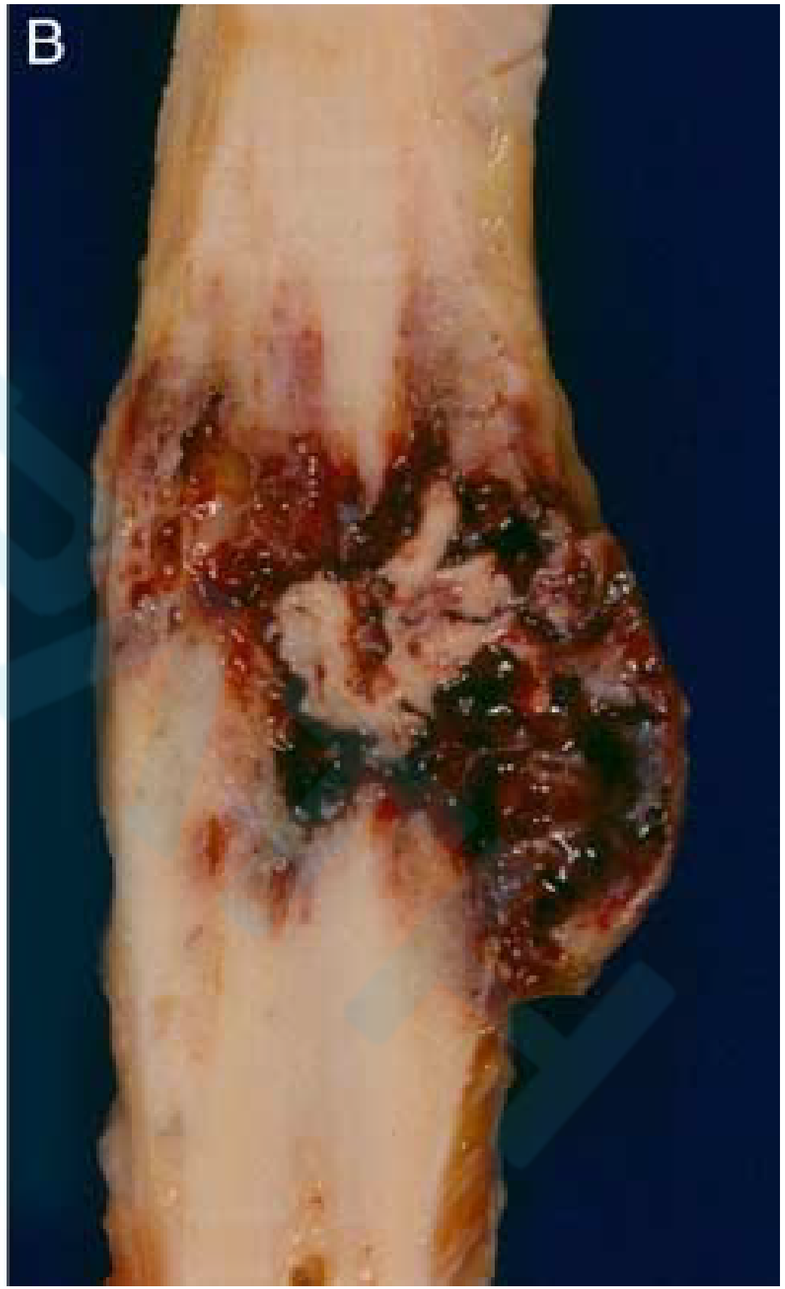




Malignant transformation and/or dedifferentiation

Case 7





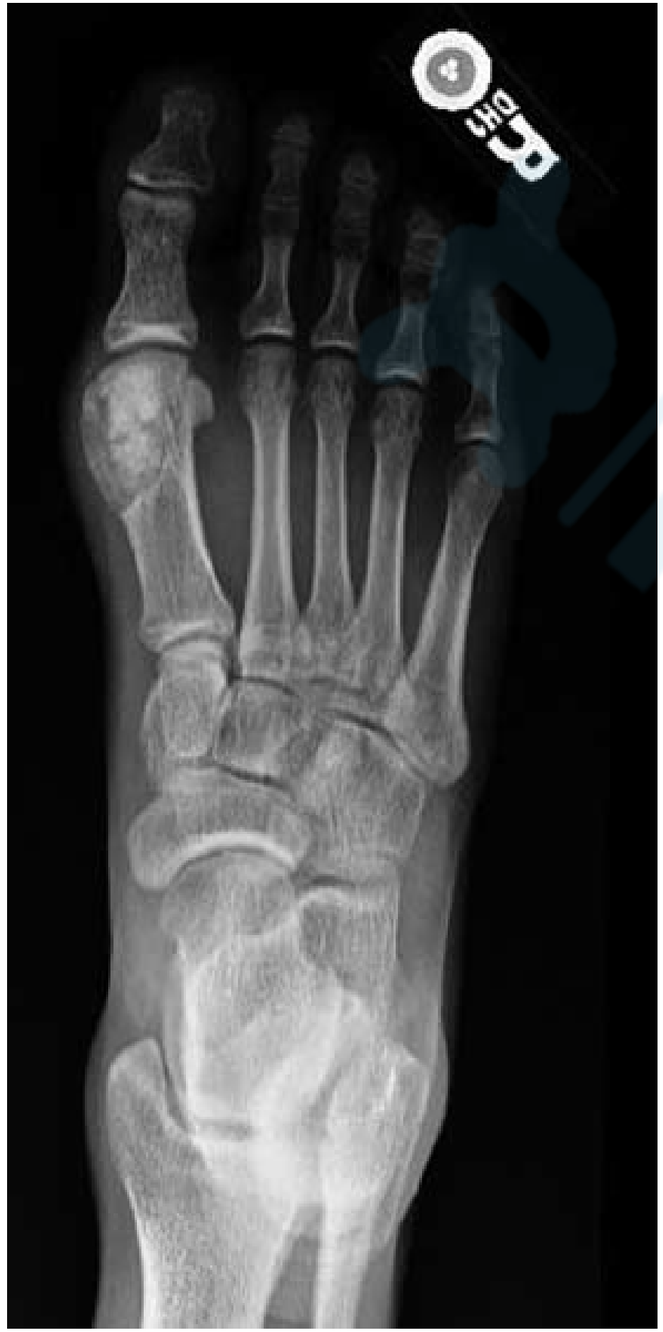
Initial Diagnosis:
Osteosarcoma

BKA performed after initial diagnostic
NED, 3 y

Expansile, largely circumscribed and intracortical lesion

Hemorrhagic to centrally sclerosing lesion

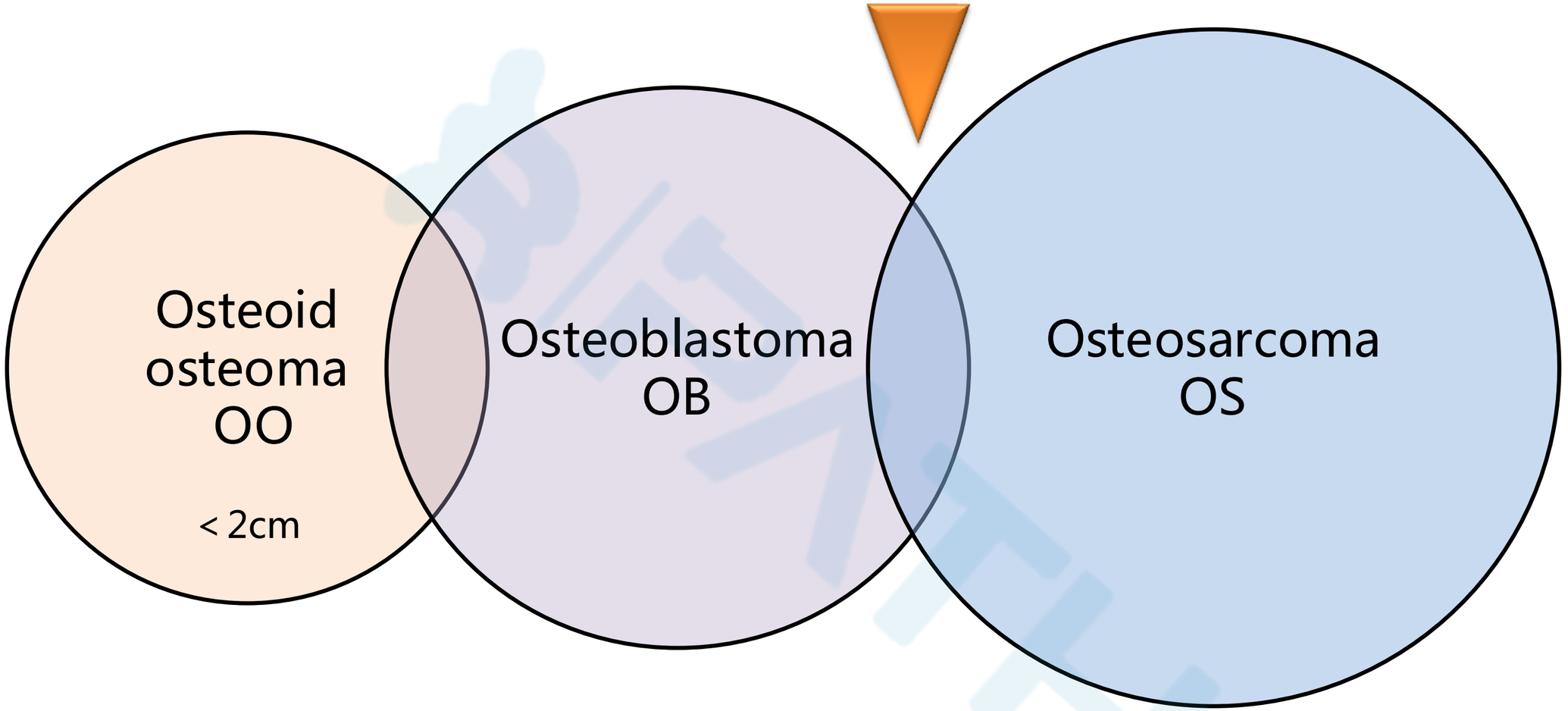
Case 8



Initial Diagnosis: Atypical sclerosing osteoblastic neoplasm
No local recurrence as of 9 mo

An expansile lesion with central sclerosis, involving the head of the first metatarsal

- With at least 12 months clinical followup for 7 of cases, the clinical course revealed a high rate of local recurrence following curettage and/or en bloc resections (80%, 4/5 cases), especially compared with conventional OB
- We report herein the clinicopathologic features of these unusual osteoblastic lesions, which we believe should be considered as borderline to low-grade malignant neoplasms



From a therapeutic and prognostic perspective, the obvious importance of distinguishing OB from OS cannot be over-emphasized

SUMMARY

- Typically arising in the distal lower extremities of adolescent and young adult males
- Radiographs generally appear as benign
- Histologic diagnosis is a dominant sclerotic matrix, with a minority of areas resembling conventional OB, without evidence of definite medullary bone permeation
- Potential for locally aggressive behavior
- Best classified by the designation "**atypical sclerosing osteoblastic neoplasm**"
- Resection with negative margins appears to be the most reasonable management option

Sclerosing Osteoblastoma

A rare type of osteoblastoma is the multifocal **sclerosing osteoblastoma**, which can be medullary or endosteal and or juxtacortical:

- Presents a multifocal growth pattern
- Roentgenological and gross features: more than one circumscribed lesion with the appearance of the central "nidus" of osteoid osteoma – "multifocal osteoid osteoma" – enclosed in a block of reactive sclerotic bone
- Histologically defined by multiple small foci of typical osteoblastoma separated by a proliferating bone and fibrous tissue
- A few may have a predominant proliferation of epithelioid cells, a nodule composed exclusively by epithelioid cells can mimic metastatic carcinoma

THANK YOU