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RADIOLOGICAL ASSESSMENT OF 78 PATIENTS OF EWING SARCOMA INTRODUCTION OF INTERESTING & UNUSUAL PRESENTATIONS

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Here is description of 78 patients studied during the period of 1356-1359(1977-1980) in the department of Radiology at the Cancer Institute of Tehran University. The goal of the study was to find out the unusual radiological presentation of Ewing sarcoma, which were jointly confirmed by histopathological studies.

History:

Ewing sarcoma is one of the well known malignant bony neoplasms, originating from the round cell of bone marrow. Luck was the first to point it out in 1866. In 1890 Hild Brand and in 1894 Marck Wald also mentioned about it. In 1916 Symmer & Vance gave its histological description.In 1921 James Ewing did a thorough reassessment of the tumour and gave new name like Endothial Myeloma and Diffuse Endothelioma.

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a) Incidence: one good study was done in Sweeden which showed that Ewing sarcoma is seen in about 10% of the primary tumour of bone.

b) Race: This tumour is usually seen in white and coloured races. Surprisingly blacks are very rarely involved in this tumour.

c) Age: More than 90% cases are under 30 and 80% out of these are in the secand decade. The youngest patient reported was 5 months age and oldest patient reported has been of 83 years age.

In our study the youngest patient was a girl of 1.5 years age and oldest patient was a man of 33 years age. The peak age in our study was 15 years which when compared with the published datas is somewhat higher. In most reports 19 years was the age of peak incidenc.

d) Sex: Male are affected more ferquently, with the male to female ratio about 1.6: 1. In our study 45 cases were males and 33 were females, the ratio comes to O:O= 1.4:1

Table one showing the age and sex incidences in our study.

Age in decades	Female	Males
first "	7	6
secand "	23	30
third "	3	9
Total	33	45

e) Trauma: In most of the reports trauma was the precedent. In our study 12 patient had a history of trauma,

9 out of them also had evidence of of pathological fracture.

f) Site of tumour: This tumour can appear any where in the bony system. 14 articles by Falk and Stephen in which 720 patients were studied, showed that 47% of these tumours were seen in the long bones and 19% were in the pelvic bone, i.e., in total 66% tumours were in long and pelvicbones. In reportfrom Tata Memorial Hospital 27% tumours were seen in femur and 17% in tibia & fibula and 9% in humerus. 50% of these tumours of long bones were seen in the midshaft area.

In our study also showed that the highest involvement i.e., 26% was seen in femur.

Table two showing the incidence related to the site of the tumour.

site of tumour in long bones	No.	Site of tumour in the other bones	No.
Femur	23	Pelvic	11
Tibia	12	Rib	4
Fibula	13	Scapula	3
Humerus	8	Metatarsal I	1
Radius	1	Calcanous	1
		Jaw	1
Total	57	Total	2

Clinical presentation:

The commonest presentation is pain and swelling. Pain is seen in 86% cases and swelling is seen in about 82% cases. Malaise may also be there.

In our study 66 patients (88%) presented with pain and 54 patients (70%) presented with swelling.Interestingly two cases had cutaneus fistulas at the site of the lesion.

Ewing sarcoma from Radiological point of view.

In 1921 Ewing declared the radiological findings as the most satisfactor finding for the diagnosis. However in 1927 Kolandy and in 1943 swe nsonmentioned that other bony lesions like osteomylities, lymphosarcoma and bony metastasis of a carcinoma can resenble to Ewing sarcoma. Lately we have found that bony lesions like Reticulum Cell sarcoma, Osteogenic sarcoma, Multiple Myeloma,Eosinophilic Granuloma and metastasis of Neuroblastoma can have findings similar to Ewing sarcoma. That is why, for a final diagnosis apart from radiological findings we have to have to take help of biopsy, staining and histopathological assessment.

A-Lond Bones:

From radiological point of view, long bone tumours can be subdivide in the following groups on the basis of site of involvement.

1- Disphyseal 2- Metaphyseal 3- Metaphyseo-epiphyse First two types can involve I) central 3 II) cortical or peripheral.

1- Diaphyseal: The commonest site of involvemnt is central diaphys The majority of which is symmetrical in

nature and midshaft in location The three forms of radiological presentation are: Lytic, Sclerotic and Mixed. Lytic lesion are mostly seen in the shaft of the bone which has a patchy or moth-eaten or faint or faint or crack-ice appearance. Sometime unusal and interesting forms are seen in which the whole bone is eroded in less than few months and all is left of the shaft on bone are a few small pioces. This can be named Vanishing' type of Ewing sarcoma. We do not know the reason for this extraordinary speed of progre ss. In the less common type i.e.; cortical diaphyseal form, the involvement is asymmetrical and involves less than a quarter of the bone. The invasior of the bony cortex appears in the form of the crater or sauce. The periosteal reaction has a sunburst or onion skin appearance and we see the spicules falling vertically on the bone. The radiological appearance is quite faint. The periostium near by the site of lesion may be torn and this creates 'Cadman Triangle'. Swelling of the surrounding soft tissue is without any ossification or calcification. Usually it is the form of flecks which mostprobably is result of escape of osteogenic cells from subperiostal area through the torn persiostium in to the surrunding soft tissue. This was observed in one case in our study.

In our study 24 cases(32.5) had diaphyseal involvement out of which 20 were of central type and 4 were of cortical type. As regards to the cellular involvement 19 were clearly lytic and one sclerotic and four mixed type. Four in this study had interesting appearance of a 'Vanishing' type. Out of these four, one is worth metioning. This was a young boy (M.T.) of 12 years age who was seen

in Sahrivar 1357 (August 1979) for a painfull swelling of a four month duration in the right arm. On X-ray it showed complete erosion of the whole shaft of the bone and only a few small pieces were left behind. Biospsy taken here in Cancer Institute was proved to be Ewing sarcoma.

(Slide number 3507-57) (Figure No. one)

2- Metaphyscal Type: In metaphyseal tumour (cen tral type) the beginin is symmetrical. Both central and peripheral types are mostly lytic, sometime sclerotic but rarely of mixed type. The periosteal reactions appear like onion skin and seldom as sunburst. There is also swelling of soft tissue. And this appearance can mimic Osteogenic sarcoma, periosteal fibrosarcoma and peripheral Chondrosarcoma.

In our study 32 cases(41.2) had metaphyseal involvement, two of sclerotic type, seven of mixed type and rest of lytic type. This when compared with studied done by others, appears be some what higher. One of the interesting patient in our study was a girl of 14 years age. (F.A.R.) who, after a trauma, started having pain in right tigh, along with swelling of the surrounding soft tissue. Total duration of the illness was five months. In the X-ray the whole of the metaphyses and the diaphyses in the proximal part of the femur had completely disapeared and only a small part of the femoral head was left in the acetabulum. Biopsy was taken and diagnosis here in The Cancer Institute was Ewing sarcoma.

(Slide number 595-58) (Figure No. two)

3- Metaphyseo-Epiphyseal type: This is a less common type. Usually it is a spill over of the malignant cells

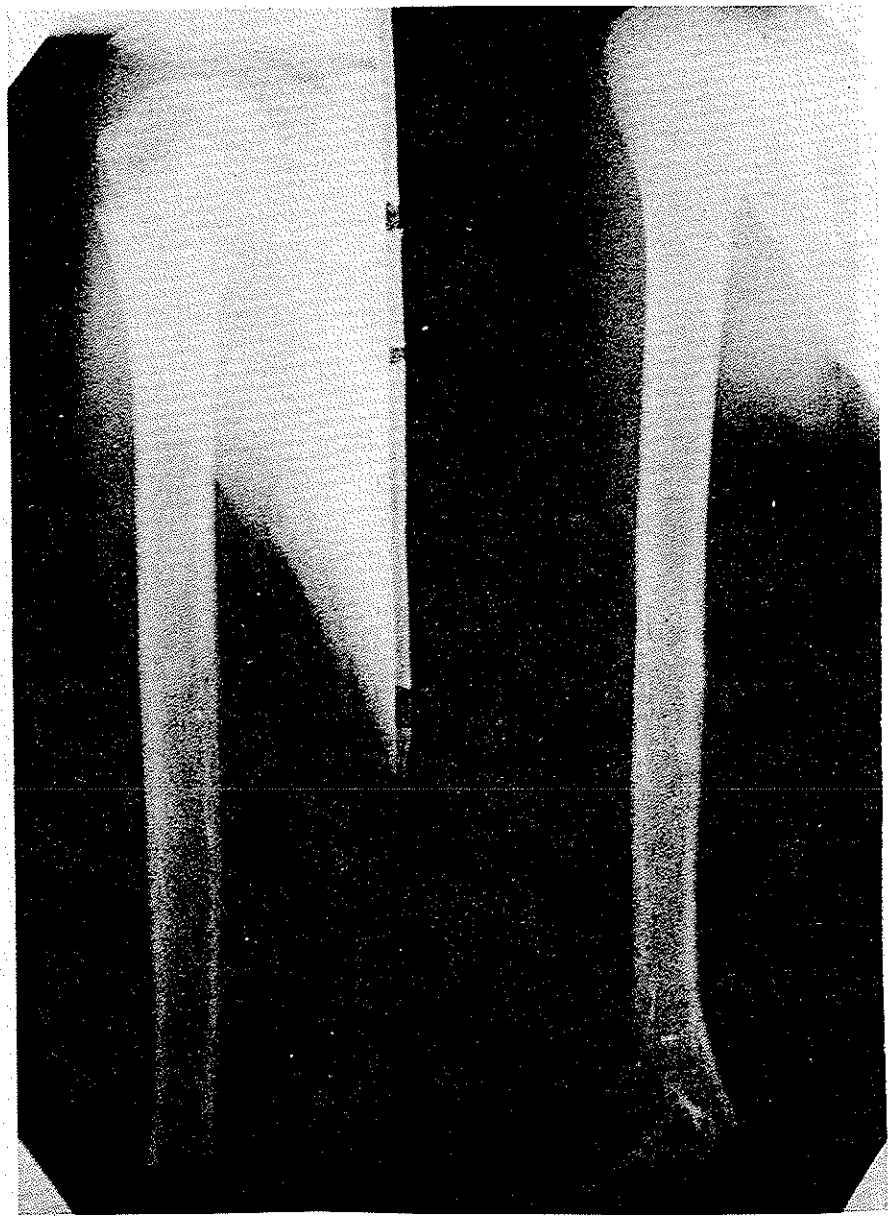


Figure No.1

Radiological Pattern No.one(M.T.)

Lytic lesion in a 12-year old boy;

a- Note the periosteal reactim in the diaphysis .

b- four months later manive osteolysis is evident.

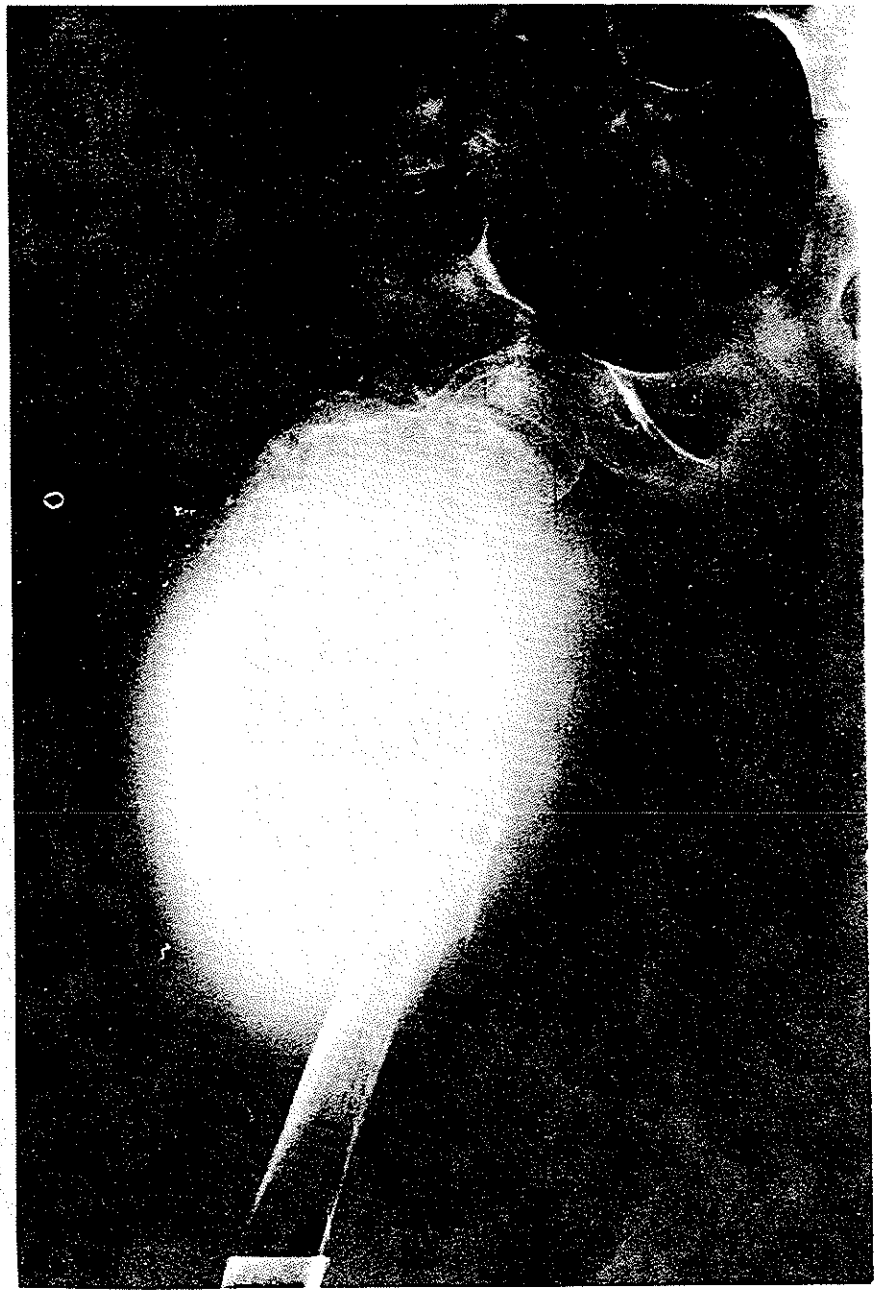


Figure No.2

Radiological pattern No. two.(F.A.R.)

Lytic lesion in a 14-year old girl;

The tumour has origirated from the diaphysis of the femur and in a months time has shown massive osteolysis.

from the nearby metaphyseas. All in all this type of the tumour is seen in less than 5% cases of Ewing sarcoma.

In our study one patient (1.3%) had metaphysee-Epiphyseal type of the tumour. This was a boy (A.M.) of 12 years age. He was seen on Azar 1358 (November 1979) for a swelling in the proximal part of right arm. X-ray showed sclero-tic and density in the metaphyseal and epiphyseal region. Spiculations and sunburst appearance was also present. There was no ossification in the surrounding soft tissue. The clinical and radiologycal diagnosis in the begening was blastic type Osteogenic sarcoma. In the Cancer Institute, biopsy was taken and the diagnosis was Ewing sarcoma.

(Slide number 6261-58) (Figure No. three)

The interesting thing was that six month later metastasis was seen in the proximal part of the left tibia and this metastasis was completely dense and blastic in nature.

B- Flat Bones:

In flat bones, the iliac and the vicinity of the sacro-iliac joint is mostly involved. Scapula is involved in less than 8%. Involvement is usually symmetrical. Mostly it is lytic in type, seldom of the mixed type and very rarely of sclerotic type. In the mixed type due to the rough walls and bony trabeculations of flat bones the lesion has an irregular foamy appearance, Ribs involvement is usually asymmetrical and the site of the lesion is commonly subaxillary and less commonly anterior and rarely in the posterior part of the chest. Periosteal reaction is either not seen and if ever seen, has onion skin appearance and is usually associated with Soft tis-

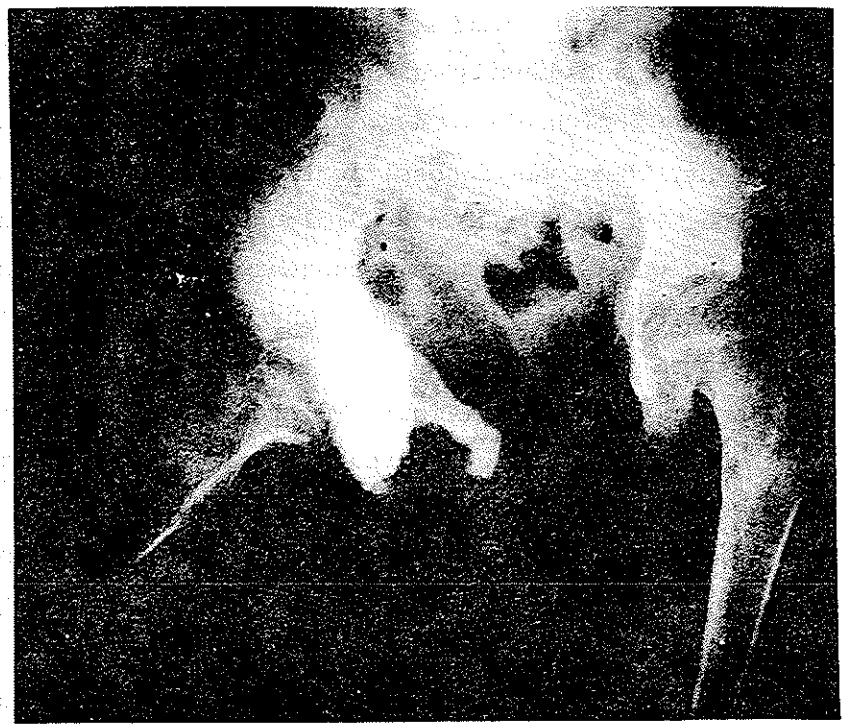
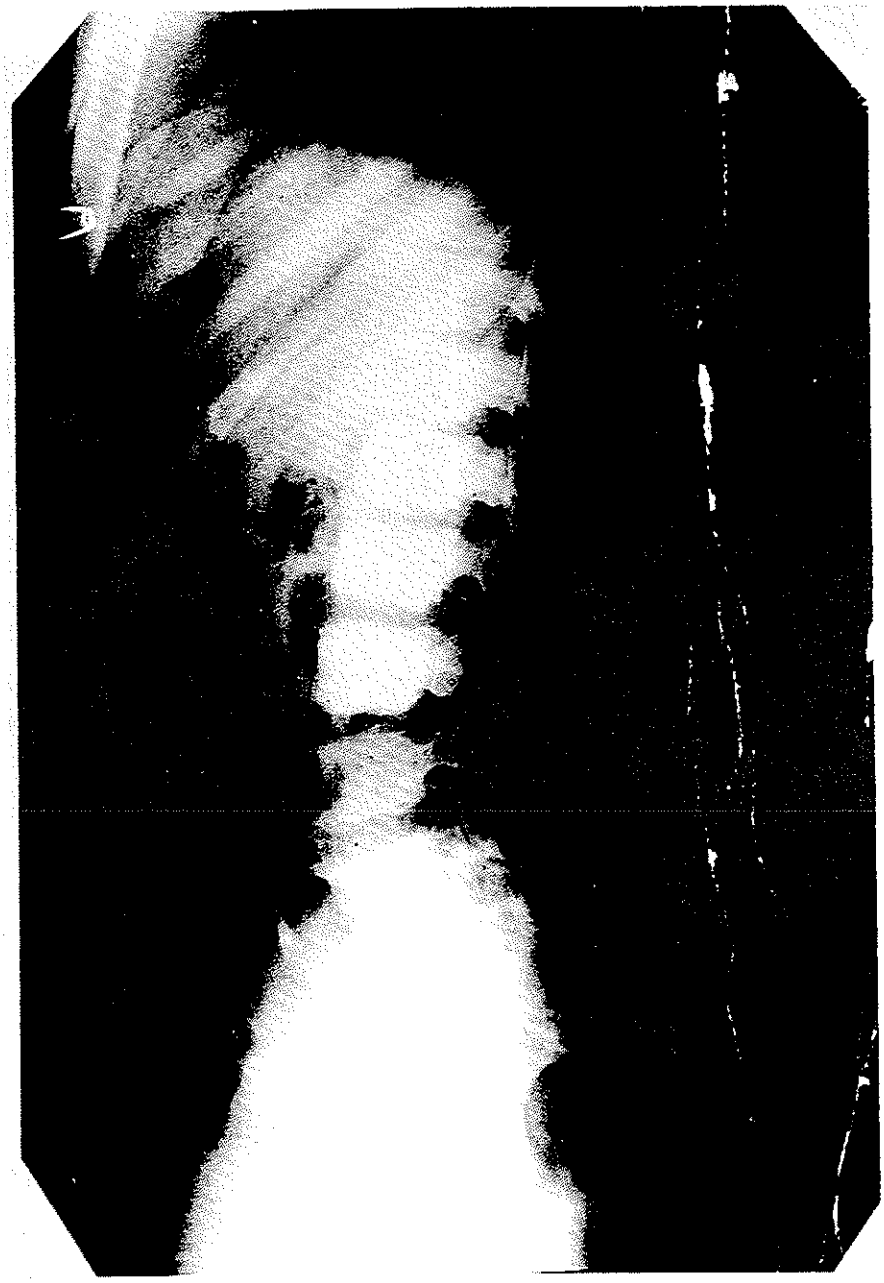


Figure No. 3

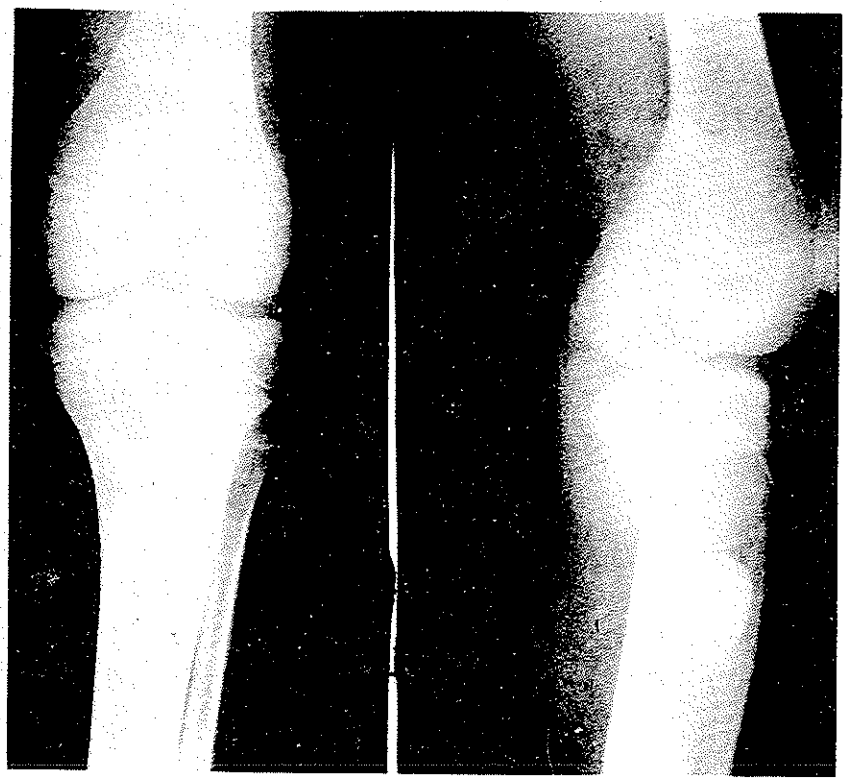
Radiological pattern NO. four.(A.S.)

Mixed-type of tumour in an 11-year old girl.

a- This showes mixed type of tumour in the right ischion.



b- After seven months an osteobastic metastasis is Visible in The lumsan spine.



b- Six months later an osteosiastic metastasis is evident in the left tibia.

sue swelling near by the ribs. This appears as extrapulmonary intrathoracic mass.

In our study 11 cases had pelvic bone tumour, six of them were in the iliac bone. One of them was sclerotic type, one mixed type and nine of lytic type. One interesting case was a girl of 11 years age (A.S.) on Aban 1358(October 1979), she was referred to Cancer Institute for swelling of right ischial bone. Before that

11-Mc Neil B.
Fluorine-18 Bone scintigraphy in children with Osteo-sarcoma and Ewing's sarcoma. Radiology 109: 627-21 Dec. 1973.

12-Mebta Y.
C.N.S. Involvement of Ewing's sarcoma
Cancer 33: 859-62 March 1974.

13- Liombart B. A.
Ultrastructure study of 2 cases of Ewing's sarcoma typical & atypical form Cancer 41(4): 1362-73 Apr.1978.

14-Potdar G. G.
Ewing's Tumour
Cli. Radiol. 22: 528-535 1971.

15-Treatment of Ewing's sarcoma
Lancet 2(8034): 391-2 20 Aug. 1965.

16-Roca A; N.
Ewing's sarcoma of maxilla and mandible.
Oral Pathology Vol. 25 No. 2 Feb. 1968.

17-Rubenstien D.A.R.
Tumour Metastasis to the Eye.
American J. of Ophthalmology. Vol. 63 No.4 1973.

18- Rosen G.
Curability of Ewing's sarcoma and Consideration for furtur therepatic trial. Cancer 41(3): 888-99 March 1978.

19- Vohra V. G.
Roentgen Manifestation in Ewing's sarcoma. A study of 156 cases. Cancer Vol. 2 NO.1 727-35 May 1967.

2- Bhansali S. K.
Ewing's sarcoma, observation of 107 cases.
The J. of Bone and Joint Surgry. Vol.-45 A No; 3 April 1963.

3- Boshgelli R. F.
Ewing's sarcoma, of the mandible, report of case.
J. Oral Surg 3(6): 473-5 Jan. 1978.

4- Buxhaum F. D.
Malignant bone tumour of lower extremity
J. Foot Surg. 15(3): 85-22 Fall 1976.

5- Dahlin C.D.
Ewng's sarcoma a cretical analisis of 165 cases.
The J. of Bone and Joint Surgery
Vol. 43-A NO. 2 March 1961.

6- Dicker, H.M.
Ewing's sarcoma of hand
The J. of Bone and Joint Surgery
Vol. 35-A No. 2 March 1971.

7- Falk S.
The Clinical and Rontgen aspect of Ewing's sarcoma
The American J. of Medical Sciences Nov. 1965.

8- Fernandez C.H.
Localzed Ewing's sarcoma Treated and results
Cancer 34: 143-8 Jul 1974.

9- Fitzer P.M.
Brain and bone scane in Primary Ewing's Sarcoma the Petrous bone. J. Neurosurg. 44(5): 603-12 May 1976.

10-Frankel R.S.
Clinical Correlation of 67 Ga. and Skeltal whole body radionuclode studies in radiography in Ewing's sarcoma.
Radiology 110; 595-663 March 1974.

Prognosis:

Ewing sarcoma is fatally progressive. The 5-years survival rate ranges is from 8 % to 28 %. At the end of one year metastasis to lung and bones is seen in about 70 % cases. In 90 % cases the cause of death is distant metastasis. Local recurrence of the tumour after radiotherapy has been described in 27 % to 33 % of the patients.

Treatment:

In our institute treatment of Ewing sarcoma is combination of radiotherapy and chemotherapy. Radiotherapy consists of irradation to the whole bone with tumour dose of 5500 to 6000 Rads on centigray (C.G.Y.) in 5 to 6 weeks by mega voltage irridation. Chemotherapy is given one course before starting irridation treatment and is continued after radiotherapy for 18 months. Surgery (amputation is generally done on complicated cases in the extremites.

Summary:

Ewing sarcoma is about 10 % of bone tumours, in the form of lytic, sclerotic and mixed type.

The lytic type erodes the whole bone in such a way that the auther would like to name it 'Vanishing' type and recognition of this radiological type is of great importance from differential point of view in the study of primary and secondry tumours of bones.

References:

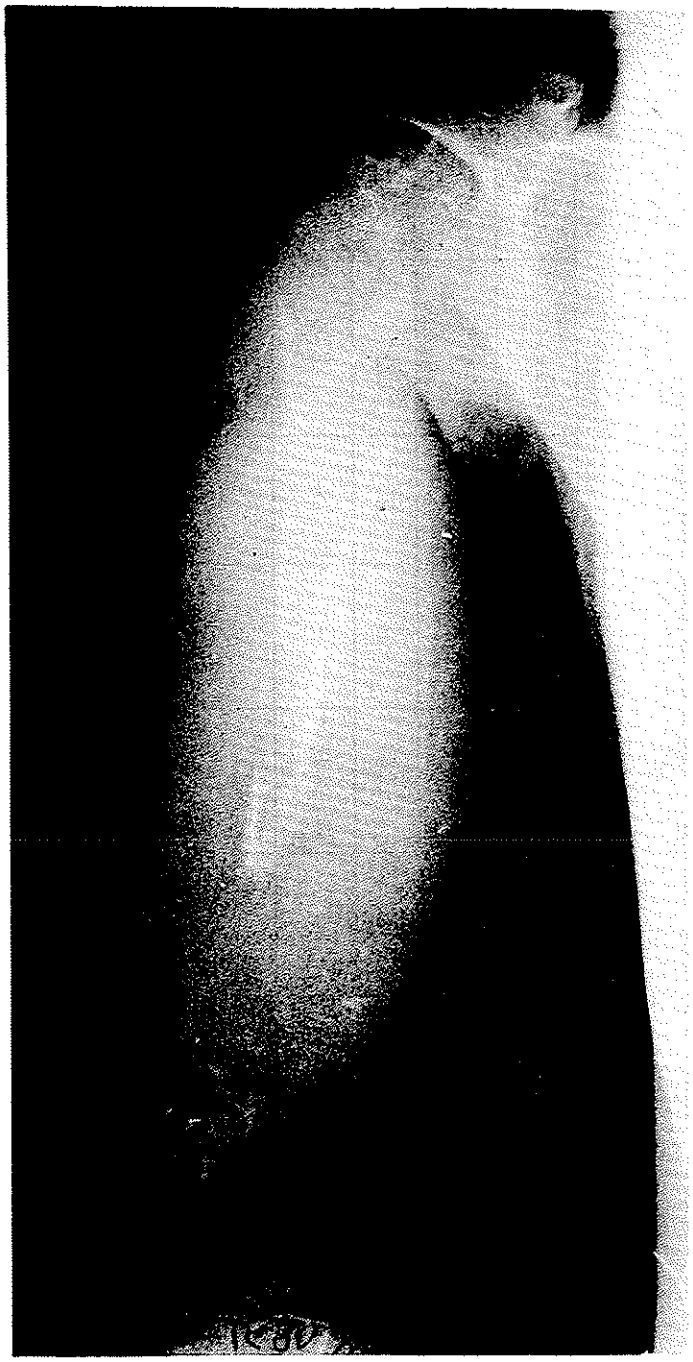
1- Aufrance, O;

Pathologic Fracture of Proximal Femur. Jama, Vol.199 No. 5 Jan. 1967.

cantly to radiography for detection of bone metastasis.

Table three showing tumour's location and radiological pathern of 78 cases.

	location	No. %	Central	Cortical	Lytic	Sclerotic	mixed
Long Bones	Diaphyseal	24(32.5)	20	4	19	1	4
	Metaphyseal	32(41.5)	24	8	23	2	7
	Metaphyseo -epiphyseal	1(1.3)	1	-	-	1	-
flat Bones	Pelvis	11(1.3)	1	-	9	1	1
	Rib	4(5.1)	-	-	4	-	-
	other bones	6(7.7)	-	-	5	-	1



she was admitted in Ahari's hospital. There the lesion on X-ray had shown sclerotic and blastic activity along with swelling of the surrounding soft tissue. Considering these findings various possibilities like chronic osteitis, osteogenic tuberclosis and blastic type osteogenic sarcoma were considered, Here in the Cancer Institute biopsy was taken and diagnosis of sclerotic type ewing sarcoma was made. Seven months later despite of radiothreapy, wide spread metastasis in the first, second and fo urth lumbervertebra were seen. Alomg with this upper part of the arm was also involve, and exactly like the primary tumour this was blastic in activity and dense in appearance. (Slide number 5874-58)
(Figure No. four.)

Four patients had involvement of the ribs,three of them had lung metastasis and it appears that the metastasis from ribs are probably more rapidly progressive on comparative basis.

Metastasis:

In more than two third of patients metastasis can be seen. The commenst site of metastasis is lung. These multiple and different size metastasis are also seen in bones and in pleura, lymph node, medastinum and C.N.S.

In our study 16 (20.5 %) had distant metastasis.14 (88 %) of them had metastasis to the lung and 11(69 %) had metastsis to skull, one of them interstingly had developed unilateral exophtalmus, two of them had metastasis to the vertebral bone. 3(19 %) of them had metastasis to lymph node and only 1(6.3 %) had metastasis to the pleura.

It is interesting that skeletal scanning is signifi-

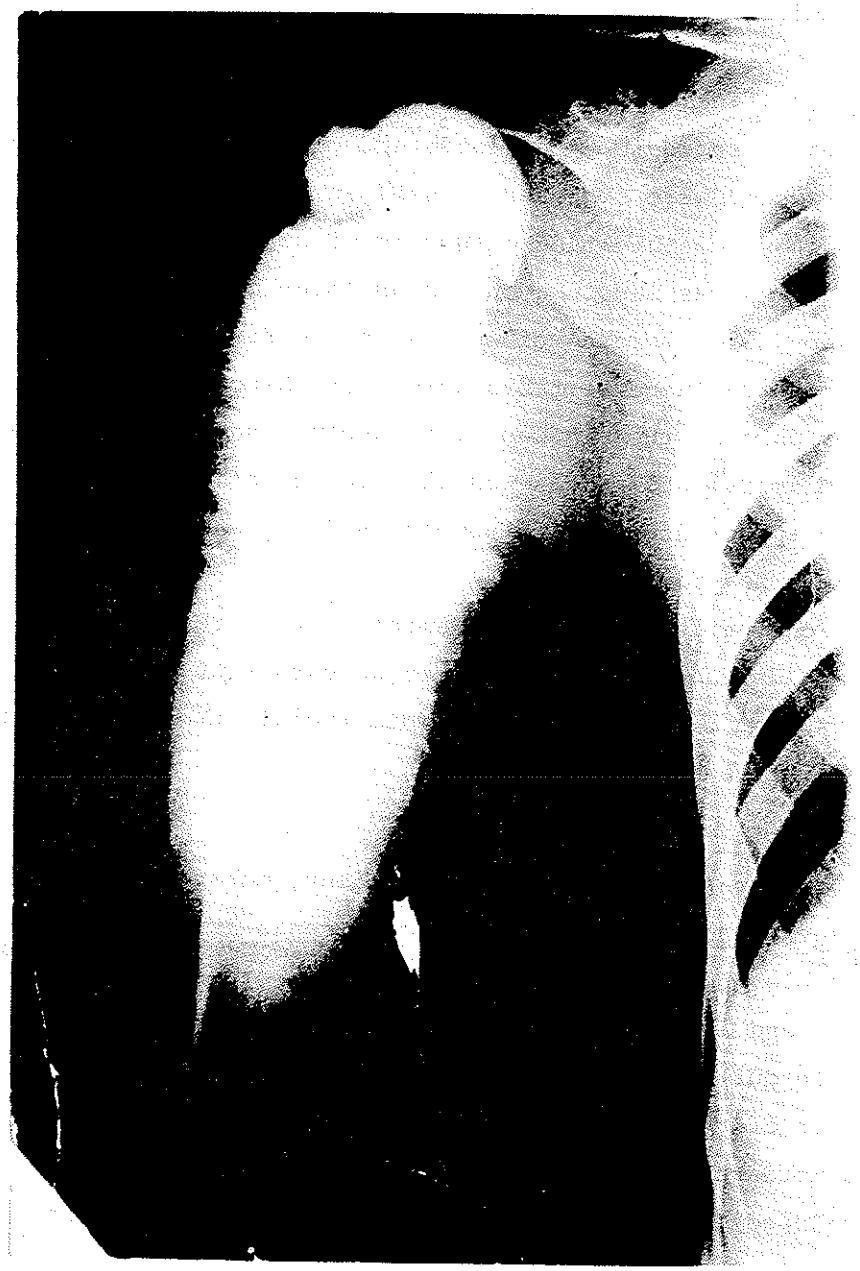


Figure NO. 4
Radiological pattern NO. three (A.M.)
Sclerotic type of tumour in a 13-year old boy.
a- This is a sclerotic type of tumour in the right humerus.