PERIOSTEAL EWING'S SARCOMA (PES). REPORT OF FIVE NEW CASES.

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Introduction :

Few cases of periosteal Ewing's sarcoma have been reported and the surgical implications of such a diagnosis have not been underlined. The aim of this study is to evaluate the actual incidence and consequence for surgeon.

Material :

All files of 135 patient treated for ewing's sarcoma of bone throughout 1982-2002 have been examined to see if they fit the diagnostic criteria and to evaluate the pronostic value and the therapeutic implications of it.

Diagnostic criteria :

The reported cases fulfilled the diagnosis criteria defined by Bator : Ewing's sarcoma of bone histologically confirmed but with a pure periosteal location without medullary extension. In our practice, the computed tomographies proved to be the most reliable exam for diagnosis. Histology of resected specimen may ignore an initial medullary involvement cured by preoperative chemotherapy. On the opposite, M.R.I. can over estimate a PES with intense inflammatory reactive tissue inside the medullary canal.

Results :

Out of 135 Ewing's sarcomas of bone of our file, only five (3.5 %) could be classified as PES. All involved the femur in the diaphyseal (2) or metaphysodiaphyseal (3) locations. According to Ennecking classification 3 tumors were graded II A ad the 2 II B. Age of the patient ranged 11 to 19. All patients were treated by resection after preoperative chemotherapy. One was irradiated..

The two first patients were not recognized as PES and were treated with wide resection interrupting the femoral continuity and skeletal reconstruction using massive prostheses; Both have still their prostheses but the orthopaedic evolution of both was complicated compelling to reoperate ; one of the patients had an acetabular wear of the hip; The second suffered of deep infection.

The three other patients were reconized as PES before the biopsy and could benefit of partial bone resection with less agressive reconstructive procedures.

With an average follow up of 9 years, all 5 patients are in first remission

Discussion and **Review of the literature** :

With these 5 new cases, only 25 cases of PES of bone have been described : 24 out of 25 patients were located on long bones. All were stage II disease. All were treated by chemotherapy and most by surgery .

PES seem to have a better prognosis than common form : 23 out 25 patients (91 %) are in first remission.

The surgical implications of Periosteal location of Ewings sarcoma must be underlined:

1°)When the diagnosis is pre biopsy suspected on CT and MRI , the biopsy should be confined to the cortical bone without medullar contamination.

2°)When the diagnosis is confirmed and the biopsy had avoided the medullar canal, a partial resection without interruption of the bone continuity must be discussed beacause such a procedure permits much easier reconstruction without massive prosthesis and avoid .

3°) the better prognosis of PES should prevent inclusion of patients in to heavy chemotherapy protocols.

Conclusion :

Periosteal Ewing's sarcoma is a rare entity that should be recognized since there is prognosis and surgical incidence.