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Pediatric chordomas: a series of 31 cases and review of the literature

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Introduction: Chordomas are rare malignant tumors of skull base and spine. Less than 5% of cases concern children, and the prognostic is essentially dependent from local recurrences. The pediatric series published in the literature are rare and generally concern few patients. We aimed to review the results of a consecutive cohort of children treated at Necker and Lariboisière Hospitals.

Methods: Thirty-one children treated between 1966 and 2012 were included. Clinical, radiological, histological and therapeutic data were collected and compared to a literature review of 250 pediatric cases.

Results: Median age was 11.8 years (3.6 – 18 years) with a male predominance (20M/11F). Intracranial and cranio-cervical junction localizations were predominant (26/31). Complete resection was obtained in 19.4% of cases in one to four steps. Twenty-nine patients had radiotherapy and four had chemotherapy. At last control, 25.8% and 37.8% of patients presented complete remission or stable disease respectively. The 15-year progression-free survival (PFS) and overall survival (OS) were 54.3% and 63% respectively. Classical histology was associated with a better OS and PFS ($p<0.016$ and $p<0.035$ respectively). There was a tendency for a better prognosis in case of complete resection, adjuvant radiotherapy and clival localization. Children less than 5 years had more aggressive forms of the disease.

Conclusion: Pediatric chordomas have a better prognosis than in adults, except for children less than 5 years. Management of pediatric chordomas has to be aggressive in order to obtain good results. However, complete resection is difficult to obtain even with staged surgeries and multiple approaches, and adjuvant treatment is essential. Identification of biomolecular markers to develop targeted therapies may improve the prognosis of these aggressive tumors.