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Skull Base Chordoma: A Case Presentation and Review of Literature

Chordome de la base du crâne : Présentation de cas et analyse de la littérature

M. O. Udoh*†, D. E. Imasogie†, D. O. Udoh‡

ABSTRACT

Chordomas are rare, aggressive, bone cancers with notochordal differentiation, a marked predilection for the axial skeleton, and higher incidence in males. Approximately 30% of chordomas occur at the skull base where they make up 0.1–0.2% of all intracranial neoplasms, and 6–16% of skull base neoplasms. Skull base chordomas typically occur between 20 and 40 years of age. We report a case of skull base chordoma in a 35 year old man, who presented with a 10 year history of symptoms, and died before treatment could be instituted. Late presentation and delay in treatment are contributing factors in the demise of this patient. The need for a high index of suspicion, early imaging and prompt referral in patients with symptoms attributable to an intracranial tumor is emphasized. *WAJM 2021; 38(2): 171–175.*

Keywords: Chordoma, Skull base neoplasm, Intracranial neoplasm, Neuropathology.

RÉSUMÉ

Les chordomes sont des cancers des os rares et agressifs, avec une différenciation notocordale, une prédilection marquée pour le squelette axial et une incidence plus élevée chez les hommes. Environ 30 % des chordomes surviennent à la base du crâne où ils représentent 0,1 à 0,2 % de tous les néoplasmes intracrâniens, et 6 à 16 % des néoplasmes de la base du crâne. Les chordomes de la base du crâne apparaissent généralement entre 20 et 40 ans. Nous rapportons un cas de chordome de la base du crâne chez un homme de 35 ans, qui présentait des symptômes depuis 10 ans et qui est mort avant que le traitement ne puisse être institué. Une présentation tardive et un retard dans le traitement sont des facteurs qui ont contribué à la mort de ce patient. On souligne la nécessité d'un indice de suspicion élevé, d'une imagerie précoce et d'une orientation rapide chez les patients dont les symptômes sont attribuables à une tumeur intracrânienne. *WAJM 2021; 38(2): 171–175.*

Mots-clés: Chordome, Néoplasme de la base du crâne, Néoplasme intracrânien, Neuropathologie.