

The Riddle of Childhood Skull Base Chordoma

Mark Yoi Sun Soo*

Senior Lecturer in Clinical Radiology, Faculty of Medicine and Health Science, University of Sydney and Visiting Medical Officer, Department of Radiology, Westmead Hospital, Westmead, NSW, Australia

***Corresponding Author:** Mark Yoi Sun Soo, Senior Lecturer in Clinical Radiology, Faculty of Medicine and Health Science, University of Sydney and Visiting Medical Officer, Department of Radiology, Westmead Hospital, Westmead, NSW, Australia.

Received: September 15, 2020; **Published:** September 30, 2020

Keywords: Chordoma; Skull Base; Paediatric

Abbreviations

SB: Skull Base; MRI: Magnetic Resonance Imaging

Skull base (SB) chordomas in children are challenging propositions in management regardless of histological grades. A dictum in the surgical-oncology fraternity states that the biological nature of chordomas is remarkable as each poses a different clinical behaviour. Moreover, data from experienced research groups have concluded some of those affected die of the disease within two years despite treatment while others survive free from disease progression for up to 20 years. To my mind, there are inherent physical structures in the patient that encourage neoplastic invasion, a situation unrelated to tumour biology.

Past experience has convincingly shown children five years of age or younger afflicted with a SB chordoma, would have a poor prognosis. This stems from its tendency towards early evolution into cellular atypia. Fortune favours the older children whose long-term prognosis is similar to that of adults. For both paediatric and adult patients, the objective of surgical treatment is gross total resection. This is seldom achievable given the lesion's invasiveness and close proximity to the lower cranial nerves and branches of the cavernous internal carotid arteries. The adverse prognostic markers therefore consist of residual tumours and post-therapy recurrences. Dedifferentiated chordomas and the poorly differentiated subtypes also belong to this list.

What truly tests our clinical acumen are lesions that extend beyond their clival boundaries into the cerebello-pontine angles or caudally to the cranio-cervical junction, impinging on the atlanto-occipital ligament. Indeed, a clival-upper cervical chordoma breaching the cranio-cervical ligaments can be life threatening. Assisted by the child's clear planes of cleavage, rapid invasion of the posterior nasopharyngeal and peri-carotid spaces is a certainty as exemplified by a 2-year-old girl with an atypical clival-cervical chordoma. She died within 48-hours of initial presentation before treatment could be instituted.

Equally serious was the case of a 10-year-old boy whose clival chordoma had caused acute headaches and visual impairment. Contrast enhanced magnetic resonance imaging (MRI) revealed a huge haemorrhagic vascular tumour extending to the petro-clival fissure. An emergency subtotal resection resulted in recovery of his sight and III and VI nerve palsies. The cause of intra-tumoural bleeding is identical to the patho-physiological change in pituitary apoplexy in which a neoplasm has outgrown its blood supply, resulting in leakage of blood through the necrotic vessel walls. This is an example of a patient's intrinsic vascular wall deficiency.

The mere size of an infant can dictate the gravity of a situation. The parents of a 7-month-old baby boy noticed he had progressive snoring and lately episodes of cyanosis, apnoeic spells and most frighteningly loss of consciousness for two to three hours. MRI confirmed a large avascular retropharyngeal clival mass. Its midline position enabled a safe transoral radical resection. The boy made a full recovery, topped by reports his tumour was a classical chordoma. Perhaps his story typifies SB chordoma's uncertain biological nature since few are entirely non-vascular.

Surgery of the skull base in the paediatric population has evolved since the start of the new millennium. Endoscopic, endonasal surgery in children aged six and less has gradually been accepted as effective and safe, particularly for mid-line lesions. A minor drawback is the lack of pneumatization of the sphenoid sinus, an anatomical landmark for trans-clival resection of chordomas. Use of neuro-navigation, real time intraoperative monitoring with MRI counts as routine. In addition, modern paediatric anaesthesia is an indispensable arm of a multidisciplinary team. Such facilities would have been in place by 2016 when a 9-year-old boy underwent radical resection for a clival-upper cervical classical chordoma. Adjuvant proton beam therapy to eradicate possible residual disease was also given. Thus, he led a life free of this dreaded condition for nine years. Unexpectedly, recurrence at the primary site necessitated a second course of proton beam therapy. Although remaining disease-free for another 12 months, he died of sudden oropharyngeal haemorrhage. The multidisciplinary team classified his case a local failure. Yet, it is conceivable a leakage of blood from the necrotic walls of the partially irradiated cervical carotid arteries had contributed to his demise.

Credit to the scientists for making inroads in understanding the biological behaviour of childhood SB chordomas. Yet, the riddle of its biological nature is at best partly resolved. Distant metastases among children five years of age or less is unpredictable. Metastatic spread to the lungs resulting in extensive and fatal pulmonary embolism was the main presentation of a 26-month-old Hispanic boy. Insidious spread to bones can concurrently happen to a few receiving chemotherapies. Even those with classical chordomas are not exempt. These are the rare and unusual cases described in the literature. Contrarily data from a recent Mayo Clinic series are more optimistic: over a mean follow-up period of 129 months only three of 35 children with predominantly SB chordomas suffered metastatic disease.

Grief-stricken by the premature passing of a close family member, my dad once wrote: "sometimes, life is so incongruous". In the context of our discussions, what he said has an element of truth.

Volume 9 Issue 10 October 2020

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