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Opinion

Paediatric Skull Base Chordoma: Does Knowledge in Neuro-Vascular Anatomy Play a Positive Role on its Prognosis?

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To a radiologist's ambition, attaining competence in neuro-vascular anatomy is invariably a challenging achievement. It relates to the complexities and configurations of the arterial supply of skull base. When it comes to surgery of a paediatric clival chordoma, a brief anatomical description is relevant. It can be regarded as a surgical road map since at times the origins of the major arteries and their anastomoses can, unexpectedly, be variable.

In any field of paediatric surgery there are fundamental and critical physical drawbacks inherent in our young patients. They have a limited blood volume. Their small body mass makes it easy to reach dangerous hypothermia. However, a multi-disciplinary neurosurgical team, equipped with intra-operative MRI and continuous neuro--sensory monitoring, should leave these to their neuroanaesthetist and the attending nurse.

The surgeon's main objectives and approach are complete tumour excision, but without damaging surrounding cranial nerves and their blood supplies. These are branches that usually originate from the cavernous internal carotid artery. They include the prominent and easily identified meningeal-hypophyseal branch and the equally important inferior lateral arteries. Careful dissection can remove the bulk of the lesion. But should this become impracticable due to potential injuries to the neighbouring cranial nerves and their respective arterial feeders, it is prudent to stop. One can resort to adjuvant radiotherapy.

The inexperienced might mistakenly regard the cavernous internal artery with its antero-superior and side branches have a symmetrical distribution. But they would be pleasantly shocked to learn up to 10% of the ophthalmic artery originates from its genu segment. This artery can on infrequent occasions anastomose with branches of the middle meningeal artery: its ligation may have serious consequences. On its way into the orbit the ophthalmic artery gives rise to the anterior and posterior ethmoidal arteries that usually run along the floor, or sometimes, below the anterior cranial fossa. A case of these arteries feeding an anterior fossa meningioma was documented. Yet, only in extreme situations whereupon a huge chordoma eroding the clivus with enormous encroachment into the anterior nasopharyngeal space shall we encounter vascular supply from the low-lying ethmoidal arteries.

The modern age of popular trans-nasal/trans-sphenoidal surgery for adult patients has resulted in a relative shortage of surgeons experienced in addressing paediatric skull base tumours. The literature reminds us of the vulnerability of a child's evolving facial bones and his or her growing jaw bones and dental eruptions. But children's well-defined soft tissue plane of cleavage helps to identify crucial landmarks. Perhaps the most nontraumatic approach is the transzygomatic in which the middle cranial fossa and the ipsilateral cavernous sinus are exposed. The three major branches of the V^{th} Nerve are also evident. Nevertheless, the more popular transmaxillary approach is safe among adults and is gradually used for children. Technically, it conforms to a LeForte One osteotomy, giving clear view and access to the upper two-thirds of the clivus. The more classic method for children is the endoscopic endonasal approach. It exposes the clivus from the posterior clinoid level to the foramen magnum. The entire tumour can be removed safely using this approach.

Complications however mild can be consequent to the safest technique. Thus, a transnasal approach for tumour excision may breach the dura and arachnoid mater. The resulting CSF (cerebral spinal fluid) leak can be as high as 20 percent. Consider the inconvenience and stress the child and family members are subjected to during repair of the leak. There is the remote possibility too of a treatable bacterial meningitis. A harder situation to cope is to find the original cranial nerve palsy has persisted and discover transient nerve palsies have appeared from minor injury to the arterial feeders.

The value of subjecting the child to post-surgical radiation therapy continues its debate: historically adjuvant radiotherapy was considered routine when surgical excision failed to remove the tumour's major portion. The school in favour of postsurgical radiotherapy has highlighted that without this mode of treatment, their patients would die from the disease very quickly-a matter of a few months to a couple of years mainly from disseminated secondary deposits. The "wait and see" school regard radiotherapy as a secondary weapon that would only be used on tumour recurrences. Moreover, radiotherapy can be relied upon should their patients need a course of re-irradiation.

Presently, with the advent of aggressive neurosurgery in centres dedicated to treat all forms of chordomas, the overall survival rate is reaching 90%--- these figures relate to the adult population. But there will be times in the near future, when the afflicted within paediatric age group can show figures to match that of the adults.

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