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# Childhood and Adolescence Chordoma: It's "Half-Forgotten" Biological Behaviours

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The biological nature of chordomas is always fascinating: not least because there has been the odd adult patient, 60 years of age, who had a sacral chordoma excised, was then afflicted with a large intrathoracic tumour of similar nature 2 years later. That he is alive and well after a second major surgery makes me re-examine the capricious nature of this tumour.

Chordomas are malignant, and exclusively so in children 5 years of age and less. We cannot be definite about the concrete reasons except histologically the tumour cells are invariably atypical with nuclear pleomorphism and necrosis. Scientists have observed some cells show sarcomatoid features although not to the extent of a classical sarcoma. In this age group dedifferentiated chordoma is not encountered unless the child has received radiotherapy at least 2 years previously. Yet genetically, there is loss of BAF47 immunoexpression correlating with tumour aggression. This genetic deficiency was reported in children in the age group of 1 to 17 years, with a peak age at 7 years. Significantly BAF47 immunoexpression loss is a poor prognostic indicator of death occurring in the first 3 years from initial diagnosis: 72% of children are thus affected in one study. Management is based on the planning and execution of a multiple disciplinary team of oncologists, paediatricians and specialists' surgeons and anaesthetists. Surgery's main aim is complete resection and if impossible to diminish tumour load so that cerebral vasculature and cranial nerves are clear of residue in the event adjuvant radiotherapy is required. Complete resection paves the way to a cure, but its difficulty poses a challenge: in one paediatric series the success rate varies from 0% to 36%.

Nonetheless some crucial factors must exist to account for this tumour's aggressive and invasive behaviour. Some ten year ago a scientist from Japan wrote an article on the "Invasiveness of Chordoma", From his deeply scientific thesis, he has proposed there are expressions of tumour-associated proteinaceous materials such as urokinase and plasminogen, that cause invasive growth of chordoma. In a similar vein, chordoma's expression of a proto-oncogene substance named "C-Met" might promote the lesion's invasive growth. Moreover, adhesions caused by tumour cells have contributed to its aggressiveness. Additionally, solid lesions such as chordomas can express cell adhesions molecule named N-Catherin that regulates the behaviour of tumour cells. A raised or positive N-Catherine is suggestive of distant metastasis.

What then are the host's defences against invasion by this malignant lesion?

With low clival lesions in young children it is technically impossible to excise them completely.

Postsurgical recurrences may extend antero-inferiorly to abut the posterior nasopharynx: to be arrested by a relatively strong anterior atlanto-occipital ligament. Once this ligament is breached tumour will spread to the posterior nasopharyngeal space, with dire consequence.

With their virtually characteristic imaging features of vertebral body erosion and collapse, chordomas of the thoracic spine are seemingly easy to treat. En-bloc spondylectomy with wide margins has been the accepted practice for decades. Yet, an intrathoracic/posterior mediastinal chordoma though uncommon, counts as a more challenging proposition. Dysphagia, difficulty in breathing, cough and retrosternal discomfort are the main symptoms. There is the odd instance of anhidrosis of both hands due to partial compression of the sympathetic chains sited along the posterior wall of in the posterior/superior mediastinum. On post-contrast MRI, a well circumscribed enhancing mass with numerous low signal septations is characteristic. It's complete removal through a posterior thoracotomy is usually successful with scant complications. Contrarily, in a rare case of an 8-year-old boy the lesion is adherent to the posterior intercostal muscles and rib cage undetected by medical imaging but only visible at surgery. There had been rare reports in which lesional adhesions and invasion of the adjacent lower lobes had extended through the diaphragm to invade the liver.

The 15 mm thick presacral fascia is regarded as a strong line of defence against invasion by sundry sacral-coccygeal lesions: chordomas and teratoid tumours being the commonest. Nonetheless, chordomatous tissues can be malleable, and may extend laterally through the sacro-sciatic notch, gaining access into the subcutaneous tissues. It's invasiveness in young children is occasionally exemplified by burrowing through the skin to present as a tender and partially haemorrhagic anterior gluteal mass.

There is much to learn of chordoma's uncertain nature. Indeed, the lesion's commonly accepted indolence in growth masks its infiltrative nature, attacking the diseased spine, its foramina, pedicles and major arterial supply. It creeps along almost silently without causing severe pain and instability until the body's defence begins to crumble. A less common group characterised by a rapid downward clinical course has been described. Two young children, both 2 years of age, had suffered from cranial and thoracic chordomas respectively They presented at casualty with severe respiratory distress due to pulmonary and pleural metastases. Both received chemotherapy but died a few weeks later from widespread metastatic disease.

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