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Excision of ganglioneuroma from skull base to aortic arch ☆

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Abstract High retropharyngeal neuroblastic tumors in children have been excised and debulked transorally or cervically, often with a covering tracheostomy. Although we and others have approached high thoracic lesions thoracoscopically, the trapdoor incision (or modification thereof) is generally reserved for cervicothoracic tumors with significant vessel encasement around the thoracic inlet. We report a case of symptomatic ganglioneuroma extending from the nasopharynx, at the level of the skull base, down to the aortic arch: macroscopic clearance was achieved via an extended trapdoor incision and without recourse to tracheostomy, transoral surgery, or transfusion.

Neuroblastoma has been reported in nasopharyngeal or cervicothoracic regions in neonates and young children; ganglioneuroma at these locations appears rarer [1-4]. Cervicothoracic neuroblastic lesions typically encase the ipsilateral vertebral and subclavian arteries and may present with Horner's syndrome. The latter is almost invariable after removal of significant tumors of the cervicothoracic sympathetic chain. Although we and others have used thoracoscopy for high thoracic lesions without significant vessel encasement, surgical approaches to encasing cervicothoracic tumors typically use varieties of trapdoor incision [5]. In contrast, more cephalad retropharyngeal tumors have been accessed transorally and/or cervically in children with similar approaches used for uncommon tumors of the adult retropharynx [3,6]. We report a case of ganglioneuroma bridging these territories that extended from skull base to aortic arch but was nevertheless amenable to macroscopic clearance via an extended trapdoor incision at a single surgery and without need for covering tracheostomy.

1. The case

This child presented around 2 years of age to the pediatric service with a lower respiratory tract infection. She had worsening feeding problems from 8 months of age and was now failing to thrive; at the time of presentation, she could not tolerate solid food and struggled with fluids such that nasogastric feeding was instituted. In addition, she also had

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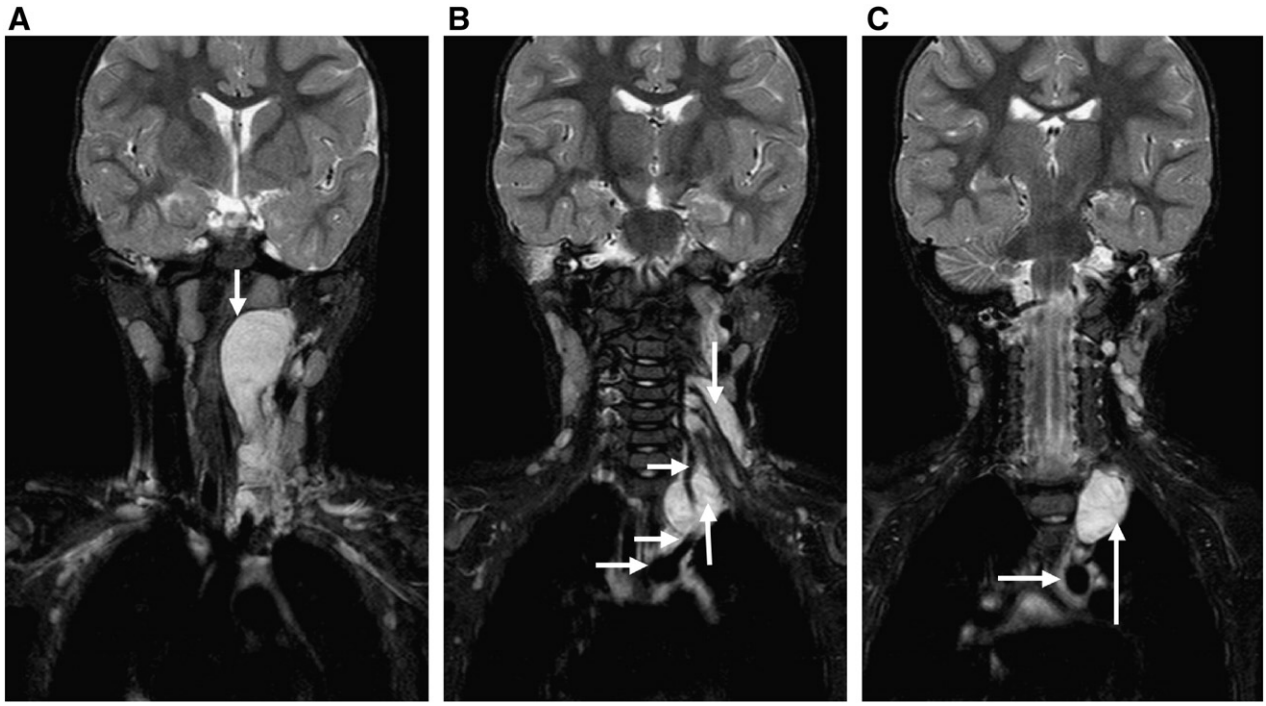

Fig. 1 Coronal magnetic resonance imaging sections from head to upper thorax showing (A) the cranial portion of the lesion (arrowed) in the nasopharynx around the level of the skull base; (B) the intermediate part of the tumor (lower vertical arrow) abutting the aortic arch (lowest horizontal arrow), encasing the left subclavian origin (middle horizontal arrow) and left vertebral artery (upper horizontal arrow) and adjacent to the brachial plexus roots (upper vertical arrow); and (C) the thoracic component of the mass (vertical arrow) and the adjacent aortic arch (horizontal arrow).

intermittent and respiratory obstructive sleep symptoms. She, like her father and older brother, was known to have neurofibromatosis type 1 with its characteristic features. Clinical examination revealed neither Horner's syndrome nor other stigmata of neuroblastic tumors. A chest x-ray incidentally revealed a left-sided apical opacity. This prompted computed tomographic scan of the area that demonstrated a cervicothoracic lesion encasing the left subclavian artery from its origin at the aortic arch, enveloping the left vertebral artery, passing behind the carotid sheath at least to the nasopharyngeal level (the highest level scanned on that occasion). After referral to medical and surgical oncology, magnetic resonance imaging was obtained to give a more complete anatomical assessment (Fig. 1) and percutaneous biopsy of the lesion was performed by interventional radiology. The biopsy specimens showed ganglioneuroma without evidence of neuroblastoma (also, urinary catecholamines were not significantly raised, and there was no hypertension). Multidisciplinary discussions involving pediatrics, oncology, pediatric surgery, otorhinolaryngology, and our craniofacial team were undertaken; although histologically benign, this lesion was producing profound dysphagia and feed aversion as well as emerging symptoms of upper airway compression/sleep apnea. Given surgical risks to cranial nerves IX, X, XI, XII, the phrenic nerve, brachial plexus, and adjacent lymphovasculature, options discussed ranged from palliation with tracheostomy and gastrostomy to formal surgical excision. Tracheostomy and gastrostomy would protect the "at-risk" airway and provide feeding access, respectively; however, the extensive underlying lesion and any concerns about occult neuroblastoma elements would remain unaddressed. Surgical excision of the lesion could avoid both tracheostomy and gastrostomy; however, surgical damage to major nerves could mandate both. The surgical oncologist (corresponding author) recommended excision if the parents were in agreement (taking the view that tracheostomy and gastrostomy could be fallback options). Debulking was rejected on the basis that with an encasing lesion, dissection had to identify and clear the critical neurovascular structures to be safe. The craniofacial surgeons also suggested the transoral approach, and covering tracheostomy, could be used for the nasopharyngeal part of the tumor.

2. The surgery

We arranged a craniofacial team to be available in the hospital if needed for a transoral approach. Fortunately, however, the tumor was removed entirely via the trapdoor approach by the pediatric surgical oncologist and assisting trainees: with a central venous line placed, a left-sided trapdoor incision was made running along the anterior border

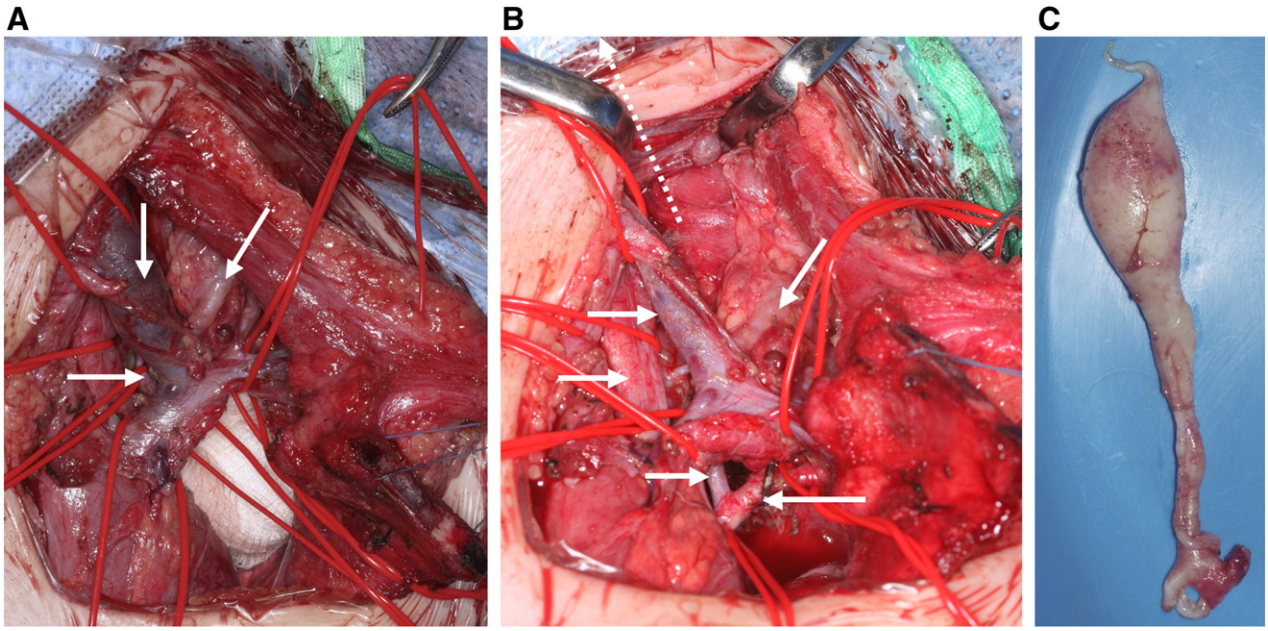

Fig. 2 Intraoperative photographs showing (A) that mobilization of the venous confluence between the left subclavian, internal jugular (vertical arrow), and innominate veins is first required to reveal tumor around the vertebral and subclavian arteries (the cleared left vertebral artery [horizontal arrow] is only just visible behind the origin of the left innominate vein). The oblique arrow marks the cleared thoracic duct (in 2B also). A swab shows the prior location of the removed thoracic part of the tumor and the adjacent aorta (see also Fig. 1C). The reflected divided manubrium is marked by a stay suture (lower right); (B) retraction of the left subclavian and innominate veins (unmarked) to reveal from bottom to top (horizontal arrows only), the left subclavian artery, left vertebral artery, left common carotid (with vagus nerve), and left internal jugular vein. The dotted arrow demonstrates the direction in which dissection of the cranial part of the tumor proceeded (its upper bound lay beyond what can be seen here); and (C) the excised cephalad part of the tumor (compare with magnetic resonance imaging in Fig. 1A) that was continuous with the cervicothoracic sympathetic chain and the lower part of the tumor (latter not shown as it was removed in blocks from around the vessels).

of sternomastoid, dividing the manubrium and extending laterally in the second intercostal space. The left internal jugular, subclavian, and innominate veins (Fig. 2A) were slooped to facilitate their retraction and allow access to the posteriorly located cervicothoracic part of the tumor and the ipsilateral vertebral and subclavian arteries (Fig. 2B). The cervicothoracic component of the tumor was first cleared from the left subclavian down to the aorta and then up along the vertebral artery toward the C6 foramen. Also expanded by tumor, the left-sided cardiac sympathetic outflow was excised. Care was taken to free and protect the phrenic nerve and thoracic duct. Moving cephalad, the tumor departed from the vertebral artery and ran within the sympathetic chain, over the brachial plexus roots and twisted behind the carotid sheath (coming to lie in the retropharynx medial to the vessels). Careful mobilization allowed the entire cranial part of the tumor (Fig. 2C) to be delivered by dissection under gentle traction via the upper cervical part of the trapdoor incision. The wound was closed with manubrial repair; a retropharyngeal vacuum drain and left pigtail chest drain were left in situ and were removed after a few days. No transfusion was required. Postoperatively, there was no evidence of tracheal compromise; a left Horner's syndrome was apparent as expected, but there was no evidence of phrenic, brachial plexus, or cranial nerve injury. Similarly introduction of oral feeds was now tolerated and not accompanied by any chylous leak. Routine convalescence was briefly interrupted by viral gastroenteritis that resolved with supportive care. Now, several months later, her obstructive sleep symptoms have completely resolved; her feeding has improved although she is still not coping normally with solids yet. Histologic examination confirmed a ganglioneuroma lacking any neuroblastoma elements.

3. Discussion

Access to cervicothoracic lesions via trapdoor incisions is well described for pediatric neuroblastic-type tumors [5]. Transoral and lateral cervical approaches have been used to access various retropharyngeal tumors in adults and more rarely children [3,6]. This lesion was exceptional in traversing both regions (and is the only one of its kind that we are aware of from our searches of PubMed and ISI Web of Science). Nevertheless, our report demonstrates that successful excision can be achieved via an extended trapdoor incision alone (Fig. 3) and without necessity for precautionary tracheostomy, transoral approach, or similar steps that can accompany major neck dissections in children and adults

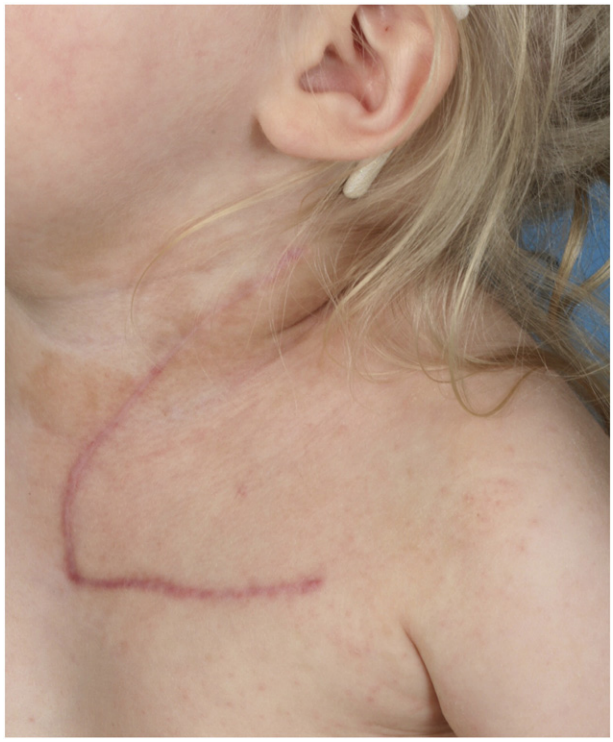

Fig. 3 Postoperative photographs showing the healing left-sided trapdoor incision: for the nasopharyngeal part of the tumor, adequate cephalad access is achievable in the young child without major extension of the incision superiorly and without measures such as temporary mandibular division.

[3,6]. A key part of safe excision was the preemptive identification and clearance of the major neural, vascular, and lymphatic structures using a tumor bivalving technique akin to the description by Kiely [7] for abdominal and pelvic neuroblastoma. The latter uses scalpel dissection along encased vessels. We use a similar approach but a different technique (modeled on that used by our cardiac surgeons for revisional operations in particular). We use low-level monopolar spray diathermy applied via a guarded spatula tip. Moreover, we used this approach for both encased vessels and encased or impinged-upon neural structures. Used at low power, this cleaves tumor from vessel, retains good hemostasis, resists inadvertent division of major structures, and like a nerve stimulator, provides warning of impending motor (but not sensory) nerves. With encasing tumors like this, such an approach allows critical anatomy to be seen and spared; therefore, "debulking" the tumor's periphery while blind to such structures should be avoided.

In addition to the unique anatomy of this tumor, the histology was also of interest. Ganglioneuromas occur at increased rates in neurofibromatosis; NF1 mutations may even predispose to ganglioneuroma in patients without clinical neurofibromatosis [8]. In this case, the excised lesion showed many histologic characteristics of neurofibroma. However, the presence of intermingled ganglia (one might say inevitably, given the location within the sympathetic chain), requires this to be classified as a ganglioneuroma.

In conclusion, we demonstrate that an extensive and symptomatic ganglioneuroma extending from the nasopharynx at the skull base down to the aortic arch is nevertheless amenable to clearance with minimal morbidity using a trapdoor incision and avoiding tracheostomy and a transoral approach. A clear strategy to identify, clear, and hence protect vital structures is essential.

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