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## Paraspinal plexiform schwannoma of unknown nerve origin: A case report

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### ABSTRACT

**INTRODUCTION AND IMPORTANCE:** Schwannomas are benign, slow-growing nerve sheath tumors of neoplastic Schwann cells. They are the most common peripheral nerve tumors in adults and are typically discovered incidentally due to their asymptomatic presentation. Despite the fact that most schwannomas are unassociated with a syndrome, their etiology is thought to be related to alterations or loss of the neurofibromatosis type two tumor suppressor gene.

**CASE PRESENTATION:** We present the case of a fifteen-year-old female who presented with a recurrent lower back/upper buttocks 9 cm mass with imaging suspicious for schwannoma. Needle biopsy revealed an S100 positive cellular schwannoma with patchy Ki-67. During surgical dissection down to the sacrum, no nerve of origin was identified.

**CLINICAL DISCUSSION:** Schwannomas have no pathognomonic findings on MRI and may occur at any location that Schwann cells are present; therefore, confirming a diagnosis relies on histopathology. Plexiform schwannomas are defined by a “network-like” intraneural growth pattern and are exceedingly rare in paediatric populations. A location distinct from the spinal canal is also very rare as schwannomas typically originate from the head and neck region.

**CONCLUSION:** Paediatric plexiform schwannomas have been rarely reported. Surgical planning relies on multiple factors such as tumor size, tumor location, pathologic features and symptomatic burden. The distinctive features of this case including an unknown nerve origin and a location outside the spinal canal provide a unique opportunity to discuss the diagnosis and management of paraspinal schwannomas and the impact on operative planning when a nerve of origin is not identified.

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### 1. Introduction and importance

Schwannomas are benign, slow-growing nerve sheath tumors originating from Schwann cells. They are the most common peripheral nerve tumors in adults [1] and usually present as solitary tumors discovered incidentally on physical exam as a lump or mass [2]. Sporadic schwannomas do not have a predilection for sex, race or age, but peak between 20–50 years of age [3]. Common clinical features of peripheral nerve tumors include a palpable mass, loss of nerve function and/or localized pain [4]. Schwannomas can arise from any central or peripheral nerve in the body with the exception of olfactory and optic nerves [5]. The areas of highest incidence include the head, neck, mediastinum, retroperitoneum, and flexor

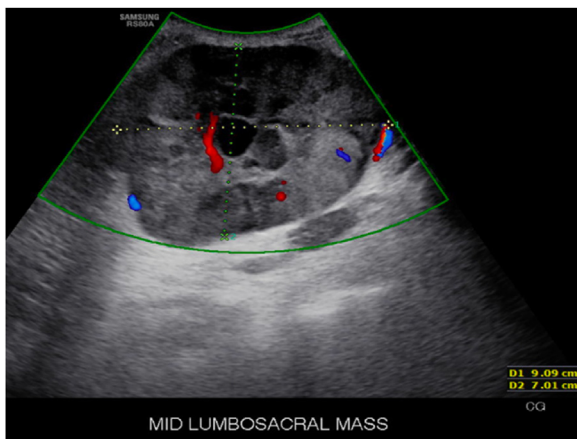
surfaces of the extremities [6–8]. The vestibulocochlear nerve (CN VIII) eight is the most common cranial nerve of origin [9].

While the etiology of Schwannomas is poorly understood, some sources suggest it relates to the alteration or loss of function in the Neurofibromatosis type two tumor suppressor gene; however, most schwannomas are unassociated with the syndrome [10]. While there is no pathognomonic imaging feature for schwannomas, computed tomography (CT) and magnetic resonance imaging (MRI) give insight into malignant features, local or distant metastasis and neural involvement. Confirming the diagnosis therefore relies on examining the tissue itself. Macroscopically, schwannomas are tan, well-circumscribed masses or macrocysts within a collagenous capsule. Histopathologic features include Verocay bodies, hyalinized vessels, and variable regions of Antoni A (densely compacted spindle fibers) and Antoni B (less dense, microcystic, hypocellular regions with macrophages and collagen) [11]. Immunohistochemical staining typically demonstrates diffuse, strong expression of pericellular type IV collagen and S-100 protein [12].

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**Fig. 1.** Ultrasound of back mass demonstrating a superficial heterogenous soft tissue mass measuring 9.09 cm × 7.01 cm.

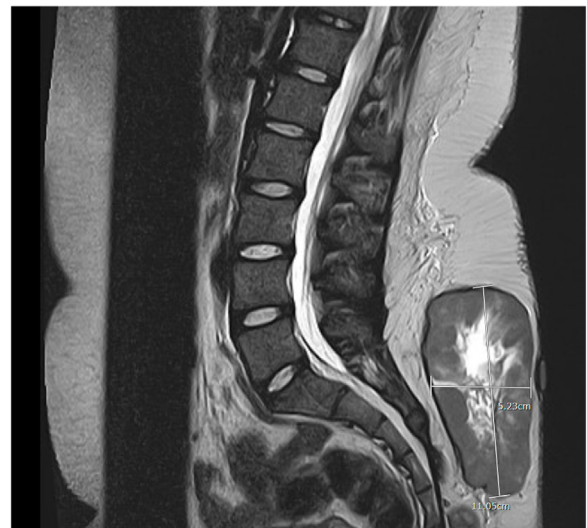
There are three specific subtypes of schwannomas (melanotic, cellular, and plexiform), each of which have unique prognosis and management. Melanotic schwannomas, as the name suggests, are distinct for the presence of dense melanin pigmentation. Unlike other schwannomas, they have the potential for malignant transformation but are exceedingly rare [11]. Cellular schwannomas are characterized by areas of high cellularity and increased mitotic activity on pathologic examination. They demonstrate a fascicular growth pattern with occasional locally destructive behavior but lack malignant potential and recur locally at a variable rate [11]. Plexiform schwannomas are named for their complex multinodular or plexiform growth pattern within neurons. They characteristically involve cutaneous or subcutaneous structures and carry a significant local recurrence rate of up to 50% despite a negligible malignant potential [13,14]. When plexiform schwannomas involve deeper anatomic structures or major peripheral nerves, they are associated with increased cellularity and mitotic activity and thus may be difficult to distinguish from malignant peripheral nerve sheath tumors (MPNST) [11]. This report has been organized in line with the SCARE guidelines [15].

## 2. Case presentation

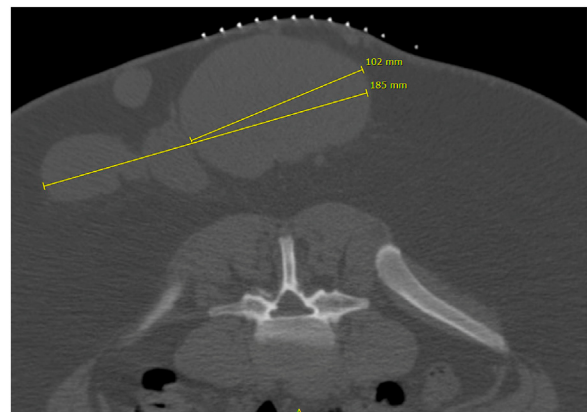
Here, we present the case of a 15-year-old female with a past medical history of obesity (body mass index 38 kg/m<sup>2</sup>) and hyperlipidemia, who presented with a recurrent mass in her lower back and upper buttocks region with possible café au lait spots identified on her upper back, arms, and legs. The patient has no family or genetic history of related disease. There is no history of drug, alcohol, smoking or psychiatric disorders in the patient or her family. The patient's mother reported the mass was surgically removed during infancy and again upon recurrence at age four years, but was unable to provide documentation or histopathologic results. Over the following years, the mass recurred with noticeable growth and symptomatic onset at six months prior to presentation, which prompted the patient to seek initial reevaluation with her paediatrician.

Preliminary imaging of ultrasound (Fig. 1) and magnetic resonance imaging (Fig. 2) are displayed below. At an outside hospital, a computerized tomography (Fig. 3) guided biopsy was performed, which revealed a cellular schwannoma with S-100 positivity and patchy Ki-67. The patient was referred to our quaternary center for surgical evaluation in the outpatient clinic.

On physical examination in the clinic, the patient was noted to have a large 15 cm soft, non-mobile mass in the lower back with overlying scar. The patient surgical planning with pre-operative



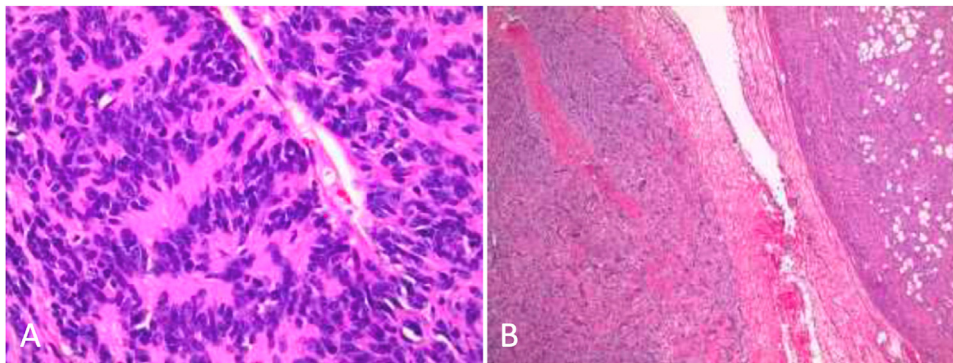
**Fig. 2.** MRI of the lower back demonstrating the large soft tissue mass with hyper-vascularity.



**Fig. 3.** CT guided biopsy demonstrating the soft tissue mass in the dorsal sacral subcutaneous tissue with multiple satellite lesions of varying dimensions.

evaluation with neurosurgery for expected neural involvement in collaboration with plastic surgery for size and cosmetic closure. After thorough discussion, patient and mother agreed to the operation and signed formal consent for the procedure and publication. The patient underwent planned surgical resection by the team of an experienced general paediatric surgeon, paediatric neurosurgeon, and paediatric plastic surgery. Intraoperative examination revealed a mass measuring 16 × 15 cm associated with multiple large feeding blood vessels and the posterior portion abutting, but not invading, the sacral fascia. A number of smaller satellite lesions were concurrently resected, some within the wall of the resection cavity. Further surgical exploration to the sacrum revealed no nerve of origin. The large wound was closed primarily without tension. Postoperatively, the patient had an uneventful course and was discharged home on postoperative day (POD) 1.

Pathologic examination (Fig. 4) of the mass measuring 15 cm in the greatest diameter showed a cystic, pink-tan, focally hemorrhagic, and variegated solid mass with central softening, containing yellow, viscous fluid consistent with a plexiform cellular schwannoma. Fibrous bands were present in the largest nodule, dividing the lesion into smaller nodules. These smaller nodules demonstrated areas of plexiform architecture with a discrete pushing border of intact fibrous capsules, variable cellular components, focal areas of necrosis and scattered mitotic activity up to three



**Fig. 4.** A. Hematoxylin and eosin staining showing variable hyper and hypo-cellularity with alternating palisading hyper and hypocellular foci, consistent with Verocay bodies (200×). B. Increased mitotic activity, focal necrosis, and fibrous bands in bundles of tumors that are intermixed with nodules, seen in plexiform schwannoma.

per ten high power fields. Immunohistochemical staining demonstrated adequate diffuse S-100 positivity and adequate patchy Ki-67 moderate activity. This pathology is most suggestive of plexiform schwannomas. No evidence of malignancy was seen.

The patient was seen for follow-up in clinic at five weeks postoperatively. She had good wound healing and happy with cosmesis.

### 3. Clinical discussion

A paraspinal mass may be caused by a wide array of pathologic conditions such as schwannoma, neurofibroma, meningioma, ependymoma, sarcoma, or other tumors arising from lymphoid, soft tissue, connective tissue, and bone [8]. Distinguishing between these diagnoses is accomplished with an array of clinical factors such as age, duration, symptom burden, family history, and physical exam, along with the histopathologic evidence.

Schwannomas may occur at any location where Schwann cells are present [9]. The rarity and wide variety of clinical manifestations may lead to delayed diagnosis and a management plan without specific protocols [2]. Preoperative imaging may provide certain clues for the diagnosis of schwannomas. On ultrasound, schwannomas are characterized by a round or elliptical cross-section with a clear border as well as an internal echo reflective of histology, which was seen in our case. On CT scan, schwannomas typically present as a well-defined and fusiform mass with relatively homogenous enhancement of contrast and internal cystic changes [5], demonstrated in our patient. While there are no pathognomonic features of schwannomas on MRI, some highly suggestive features include a well-circumscribed elliptical or spherical tumor, low T1 and high T2 signal, homogeneous contrast enhancement and a demonstrated nerve of origin or exit [2]. Schwannomas also characteristically demonstrate extra-fascicular growth which is consistent with the subcutaneous location of the mass in our case.

Surgical planning relies on multiple factors such as tumor size, tumor location, pathologic features and symptomatic burden. Total surgical resection is the gold standard for treatment of spinal schwannomas [16], especially if the mass can be safely isolated from nearby neurovascular structures, as with our case. Identifying a nerve of origin enhances operative planning and patient counseling.

For example, determining the nerve of origin for vestibular schwannoma may help predict the success of hearing preservation postoperatively [17]. Preoperative, in-office lidocaine injections have been used to enable identification of the nerve of origin of schwannomas by inducing a temporary nerve deficit [18]. Because schwannomas are well encapsulated and often displace nerve fascicles, it is generally believed that separation of the tumor from the nerve of origin can be done without producing a neurologic deficit [19]. Previous studies have demonstrated no neurologic complica-

tions postoperatively when the tumor was located in unidentified terminal branches in skin or muscle [20]. In the present case, careful surgical dissection to the sacrum did not show a connection to the spinal canal or isolate a nerve of origin, but the mass most likely originated from a nerve based off tissue biopsy.

Characteristic gross pathologic findings of schwannomas include a fibrous capsule and hyalinized vessels [11]. Recurrence of conventional spinal schwannomas is reported in less than 5% of patients who undergo surgery [16]. Histopathologic hallmarks that aid in the diagnosis of schwannomas include varying portions of a fascicular-type tissue (Antoni A) and a reticular-type tissue (Antoni B). Schwannomas may be reliably differentiated from similar looking lesions like histiocytomas and leiomyomas by the presence of laminin. This is a glycoprotein produced by Schwann cells present in Antoni A tissue due to increased basement membrane structures [8,21]. The overexpression of laminins also explains the pathogenesis of Verocay bodies, which are diagnostic of Schwannomas [21]. In our case, classic histopathologic findings combined with S-100 and Ki-positivity confirmed the diagnosis of paraspinal plexiform schwannoma.

### 4. Conclusion

Paediatric plexiform schwannomas have been rarely reported. This case of plexiform schwannoma is especially unique given the location outside the spinal canal without an identified nerve of origin intraoperatively. These features provided an opportunity to discuss the diagnosis and management of paraspinal schwannomas and the impact on operative planning when a nerve of origin is not identified.

### Declaration of Competing Interest

The authors report no declarations of interest.

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### Ethical approval

This is exempt from ethical approval.

### Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy

of the written consent is available for review by the Editor-in-Chief of this journal on request.

### Author contribution

Spencer Pace: study concept, data interpretation, writing the paper.

Marla A. Sacks: study concept, data collection/analysis/interpretation, writing the paper, manuscript editing.

Tanya Minasian: study concept, surgical participation, manuscript review.

Asra Hashmi: study concept, surgical participation, manuscript review.

Faraz A. Khan: study concept/design, surgical participation, manuscript review/editing.

### Registration of research studies

Not applicable.

### Guarantor

Marla A. Sacks & Faraz A. Khan.

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