

Peer Review File

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Reviewer A

1. “To date, only 39 cases of intradural extramedullary capillary hemangiomas have been reported in the English literature, and all of these cases have been described at the lumbar and thoracic spinal levels.” Please change this sentence to (a).

a. Note that intradural extramedullary capillary hemangiomas in the cauda equina are rare (only 16 cases), and rarely seen in the paediatric population (first case reported in your reference paper number 4). You can be more precise in quoting this number, given its relevance to your paper, to highlight how rare this tumour is in the cauda equina and your select population.

b. Can consider including a table of cases (of IDEM capillary haemangiomas), with classification based on age and location.

Reply 1:

a. Thank you for pointing out the rarity of this pathology, specifically when located at the cauda equina. After further investigation of the literature, we found that 20 cases of IDEM capillary hemangiomas in the cauda equina have been reported. Of those, only 1 patient was pediatric. We updated our text to reflect this information as advised.(see Page 3 under Introduction & Page 4 under Discussion.)

b. While we recognize the potential value of including a table of cases to illustrate the spatial and age-related distribution of this uncommon pathology, we wish to highlight that the paper we are adding into the references (Hughes et al) already features a comparable table. Consequently, the inclusion of our own case in this table may not contribute substantially to altering the information presented in the authors' existing table.

Changes in text:

“Only 20 cases have been previously reported in the literature, with the sole pediatric case being in a 17-year-old. ~~Roughly 40 cases have been reported in the literature, with the youngest being in a 17 year old.~~”

“Only 21 cases of intradural extramedullary capillary hemangiomas of the cauda equina have been reported, including our own.¹ When considering the entire spine, approximately 40 total cases of these tumors have been reported.² Additionally, our case is just the second pediatric instance to be described.”

2. Under discussion- “Intradural capillary hemangioma is a rare benign neoplasm that has been reported about 40 times in adults. Most frequently, this has been found in the thoracic or lumbar spine, with only one reported case in the cervical spine”

- a. Note the repetition of this sentence in introduction and discussion sections, could consider rewording.
- b. Please change this sentence too to make your conclusion more relevant, as mentioned in (1).

Reply 2:

a.b. Thank you for pointing out the repetition here. We updated this as advised and added further specification. Please see changes that were made in response to Comment 1.

Changes in text:

“Only 21 cases of intradural extramedullary capillary hemangiomas of the cauda equina have been reported, including our own.¹ When considering the entire spine, approximately 40 total cases of these tumors have been reported.² Additionally, our case is just the second pediatric instance to be described.”

3. Not much discussion about the imaging features of intradural extramedullary capillary hemangiomas.
 - a. Can have one paragraph on the imaging features and differentials / mimics (especially in the paediatric population).
 - b. Note there are only MRI pictures. How was it diagnosed initially? Was there any diagnostic difficulty? Any radiographs or CTs? Any vascular voids?
 - c. Figures- can include radiographs/CTs, also the pre-contrast T1 image. Any GRE/SWI images?
 - d. Figure 1.- 4 pictures with no details of the imaging characteristics- for example, you showed T2w images, is the lesion T2w isointense or hyperintense?

Reply 3:

- a. Thank you for this suggestion. We incorporated a paragraph in the discussion section touching on these points as advised. To our knowledge, there is no significant imaging differences in the pediatric population.
- b. The patient did not have XR or CT scan prior to the MRI. Based on the patient's symptoms, only an MRI was obtained which led to the discovery of the lesion.
- c. We have added a sagittal and axial T1 sequence showing the isointense characteristic of this lesion. There were no GRE/SWI sequences obtained.
- d. We have updated the text and figure legend to include details of the imaging characteristics.

Changes in text:

“Capillary hemangiomas are typically described on MR imaging as isointense on T1-weighted images, hyperintense on T2-weighted images, and homogeneously enhancing on post-contrast imaging, similar to our case. When occurring in the CNS, these tumors are usually well defined intradural extramedullary lesions.⁶ The differential diagnosis for masses with these radiographic characteristics include meningiomas most commonly, as well as capillary hemangiomas, metastatic lesions, lymphomas,

paragangliomas, and ependymomas. Other vascular pathologies should also be included in the differential such as cavernous hemangiomas, hemangioblastomas, and angiomyolipomas.^{3,6} “

“Figure 1:

Pre-operative MRI showing a well-circumscribed lesion that is isointense on T1 (A, D), T2 hyperintense (C, F), and homogenously enhancing on T1 post contrast (B, E) intradural, extramedullary lesion centered at L2 with severe spinal canal stenosis and compression of the cauda equina. There is also T2 flow void indicating vascularity of the lesion.”

In figure legend:

“Figure 1:

Pre-operative MRI showing a well-circumscribed homogenously enhancing and T2 hyperintense intradural, extramedullary lesion centered at L2 with severe spinal canal stenosis and compression of the cauda equina. There is also T2 flow void indicating vascularity of the lesion.”

4. Management-

- a. Was this a low flow malformation? Would ablation or minimally invasive techniques been useful? Why open surgery? Were conservative management techniques employed before surgery (eg. analgesia or steroids)? Any post-op radiation?
- b. Can have a description of the general management techniques.

Reply 4:

- a. The patient did not undergo an angiogram so it is difficult to characterize the flow of the lesion. Intraoperatively there was a feeding vessel that was coagulated at the time and bleeding of the lesion was virtually nonexistent after this. We have added a note about this in the manuscript in the operative technique section. Open surgery was performed to ensure an adequate decompression and resection of the lesion, and additionally perform the laminoplasty, which would unable to be performed with a minimally invasive approach. Given the patient was becoming symptomatic and the severity of the cauda equina compression, conservative management was not attempted in this patient. They did not undergo any post-op radiation. We have added a paragraph with a description of general clinical information and management techniques and added a reference that serves as a general review of this pathology (Panero et al).
- b. See above

Changes in text:

“The dura was opened along the midline and a classically described mud-brown/purplish mass was visualized arising from a single, thickened nerve root. (Figure 2) There was also a feeding vessel that was identified and coagulated during the

dissection. Intraoperative electrophysiological monitoring was employed. The entering and exiting roots from the mass were stimulated up to 1.5mA with only a very weak anal sphincter response. Surrounding nerve roots briskly stimulated at 0.5mA. The decision was made to amputate this nerve root above and below the tumor in order to remove the mass en bloc. After careful arachnoid dissection, the tumor was removed in one piece. (Figure 3) In general, hemostasis was not difficult to achieve despite the vascular nature of the lesion, most likely due to the coagulation of the feeding vessel prior.”

“Patients typically present with symptoms of other compressive lesions, including back pain, lower limb pain and numbness, lower limb weakness, and paresthesias that are typically progressive over time, ^{1,4,5,6,7,8} however, there have been reports of acute onset of symptoms due to bleeding of a capillary hemangioma.³ The most common age range for patients are between 40 and 60 years ⁷ Due to the risk of rupturing that could cause acute neurologic decline, total resection, if possible, is treatment of choice ^{1,4,5,6,7,8} Total resection also decreases the likelihood of recurrence necessitating further surgery. At this time there is no role for radiation.”

5. Laminoplasty vs laminectomy in the paediatric population

- a. Is there a significant benefit to use of laminoplasty? Any figures or quantitative assessment of the reduction in morbidity or deformities?
- b. As compared to the 17 year old’s surgical management, why was laminoplasty considered instead? Was the 17 year old’s case a good guide to management?
- c. Note that the focus of your paper was mainly on neural preservation. How about preservation of the vascular structures (given the propensity of this entity to bleed)? Was there any consideration of pre-op embolization?

Reply 5:

- a. We added a paper by McGirt et al which found that laminoplasty was associated with decreased progression of spinal deformity requiring fusion when compared to laminectomy for the resection of spinal cord tumors. This paper along with surgeon experience helped guide the decision to perform laminoplasty for our patient.
- b. For reasons described above we did not use the 17-year old’s case as a guide to management and elected to perform laminoplasty instead of laminectomy.
- c. We did not consider pre-op embolization as we did not anticipate a large amount of blood loss from this lesion as the plan was to remove en bloc. We did encounter a feeding vessel which was coagulated intraoperatively and we have added this to our manuscript (see comment 4 for the additional text).

Changes in text:

“Further in the pediatric population, laminoplasty was found to be associated with decreased rate of progressive spinal deformity requiring surgical correction.¹⁰” (See last paragraph under discussion section)

6. References

a. Generally ok, can include a general reference paper too, for the pathology, imaging features and management techniques (eg. why is total resection unequivocally the first line management).

- a. We have added a reference (Panero et al) that serves as a general overview of pathology, imaging, and management techniques, as well as details previous cases.

Reviewer B

1. Lines 62 and 161, please introduce the abbreviations ‘MRI’ and ‘CNS’ in their first appearance in the text.

60 Case Description:

61 A 14-year-old female presented with two-month history of low back pain with bilateral leg pain

62 and numbness. MRI revealed an L2 intradural, extramedullary mass causing severe spinal canal

63 stenosis. Patient underwent laminoplasty for resection of an intradural tumor. Post operatively,

159 Capillary hemangiomas are typically described on MR imaging as isointense on T1-weighted

160 images, hyperintense on T2-weighted images, and homogeneously enhancing on post-contrast

161 imaging, similar to our case. When occurring in the CNS, these tumors are usually well defined

162 intradural extramedullary lesions.⁶ The differential diagnosis for masses with these radiographic

Reply 1:

The abbreviations were introduced appropriately. Thank you for making the authors aware of this mistake.

Changes in text:

“A 14-year-old female presented with two-month history of low back pain with bilateral leg pain and numbness. Magnetic resonance imaging (MRI) revealed an L2 well-defined homogenous contrast-enhancing intradural, extramedullary mass causing severe spinal canal stenosis.”

“Magnetic resonance imaging (MRI) revealed a well-defined homogenous gadolinium-enhancing intradural, extramedullary mass centered at L2 causing severe spinal canal stenosis and posterior displacement of the spinal nerve roots. There was also a T2 flow void, indicating vascularity of the lesion (Figure 1).”

“Capillary hemangiomas are typically described on MRI as isointense on T1-weighted images, hyperintense on T2-weighted images, and homogeneously enhancing on post-contrast imaging, similar to our case. When occurring in the central nervous system (CNS), these tumors are usually well defined intradural extramedullary lesions (6).”

2. Reference(s) should cite to the introduction section. Please check it.

Reply 2:

References were included in the introduction section appropriately.

Changes in text:

“Introduction

Intradural extramedullary capillary hemangiomas of the cauda equina are exceedingly rare malformations arising from the endothelial cells of the nervous system vasculature (1). These are benign tumors owing their pathologic propensity to mass effect leading to pain and neurologic deficits. Only 20 cases have been previously reported in the literature, with the sole pediatric case being in a 17-year-old (1-4). Here we report a case of an intradural, extramedullary capillary hemangioma of the cauda equina in a 14-year-old. This article is presented in accordance with the CARE reporting checklist.”

3. In general, the submission of a Case Report should be accompanied by written consent from the subject (or their parent/guardian) before publication; this is particularly important where photographs are to be used or in cases where the unique nature of the incident being reported makes it possible for the patient to be identified.

Describe this information in **both the “Case Presentation” section of Main Text and the “Ethical Statement” section of Footnote.**

*- Suggested wording: “All procedures performed in this study were in accordance with the ethical standards of the institutional and/or national research committee(s) and with the Helsinki Declaration (as revised in 2013). Written informed consent was obtained from the *** for publication of this case report and accompanying images. A copy of the written consent is available for review by the editorial office of this journal.”*

Case reports should include an ethical statement indicating whether written consent has been obtained from the subject (or their parent/guardian). See the “Ethical Statement” section for details.

Reply 3:

The suggested wording was used for the ethical statement in both the case presentation section and in the footnote. We describe how we were unable to obtain written consent for publication after multiple attempts to contact the patient and family. We also highlight how we craft the manuscript in a way that avoids inclusion of identifying patient information.

Changes in text:

“Ethical Statement: The authors are accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved. All procedures performed in this study were in accordance with the ethical standards of the institutional and/or national research committee(s) and with the Helsinki Declaration (as revised in 2013). Written informed consent for publication of this case report and accompanying images was unable to be obtained from the patient or guardians after all possible attempts were made on several occasions. The patient information provided in this report is written to ensure anonymity. Likewise, the accompanying visuals, such as MRI scans, pathology slides, and close up intra-operative photos, are devoid of any identifying characteristics.”

4. Figures

1) Each figure must be saved and submitted as a separate file. The preferred format for figures is JPG, TIFF, or PDF format. And, please check the figure legends which are inserted after the reference list, to see if they are correct.

2) Figure 5 legend and line 136, please check the time in the main text and figure legend. Should it be 3 months or 4 months?

Figure 5. Post-operative MRI at 4 months.

Left: T2 sagittal without contrast demonstrates no residual tumor with metal artifact seen due to laminoplasty repair screws; Right: Axial T2 without contrast demonstrates decompressed nerves with no residual tumor.

136 Post operatively, the patient awoke with no deficits and resolved leg pain. A 3 month post-
137 operative MRI revealed no tumor recurrence and fully healed lamina. (Figure 5) She was cleared
138 to resume competitive sports.

3) Any abbreviations used in figures and tables or their description should be defined in a footnote beneath each corresponding table/figure. Even if they were explained in the main text, full terms must be presented again in the corresponding figures and tables, so that figures and tables can be read on their own.

Reply 4:

1. The figures were downloaded as PDFs after the figure legends were checked.
2. Thank you for pointing out this mistake. The figure title should read “Post-operative MRI at 3 months”. The figure legend was changed accordingly
3. Abbreviations were added as a footnote beneath the corresponding figure.

Changes in text:

“Figure 1: Pre-operative

MRI showing a well-circumscribed lesion that is isointense on T1 (A, D), T2 hyperintense (C, F), and homogenously enhancing on T1 post contrast (B, E) intradural, extramedullary lesion centered at L2 with severe spinal canal stenosis and compression

of the cauda equina. There is also T2 flow void indicating vascularity of the lesion. MRI = magnetic resonance imaging

Figure 2. Intra-operative Photographs

Top left: Inferior aspect of tumor with nerve roots draped over tumor and thickened nerve root exiting tumor; Top right: Superior aspect of tumor with nerve root entering tumor; Bottom right: Middle aspect of tumor surrounded by nerve roots; Bottom left: Dissected tumor being removed.

Figure 3. Gross Pathology

2 cm tumor specimen with entering and exiting nerve root.

Left: Dorsal aspect of the tumor; Right: Ventral aspect of the tumor.

Figure 4. Intra-operative Imaging Post-resection

Top: Decompressed cauda equina; Bottom: Watertight dura closure with prolene suture.

Figure 5. Post-operative MRI at 3 months

Left: T2 sagittal without contrast demonstrates no residual tumor with metal artifact seen due to laminoplasty repair screws; Right: Axial T2 without contrast demonstrates decompressed nerves with no residual tumor. MRI = magnetic resonance imaging

Figure 6. Microscopic pathology

Right: The neoplasm is composed of a cellular proliferation of round to spindled cells with minimal pleomorphism with no significant mitoses or apoptotic bodies seen (Hematoxylin and eosin, 200x magnification); Left: An immunohistochemical stain for CD34 demonstrates strong positivity, highlighting the vascular spaces and the background proliferating cell population (CD34, 100x magnification).”