

Infantile Calcifying Pseudoneoplasm of the Neuraxis (CAPNON) at the Craniocervical Junction with Postoperative Craniocervical Instability: A Case Report

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Case Report

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Abstract

Calcifying pseudoneoplasm of the neuraxis (CAPNON) is a rare, non-neoplastic fibro-osseous lesion of the central nervous system. We report the youngest patient with CAPNON documented to date — a one-year-old male, born extremely prematurely at 27 + 6 weeks' gestation, presenting with progressive craniocervical junction stenosis causing cervical myelopathy, apnoea, and respiratory compromise. Despite a computed tomography appearance suggesting dense calcification, the lesion was intraoperatively found to consist of thick, opaque bright-white fluid, representing an atypical gross finding not previously described. C1–C3 laminectomy with partial debulking achieved significant decompression. Histopathology confirmed CAPNON. Postoperatively, craniocervical instability was managed with prolonged rigid cervical immobilisation, avoiding surgical fusion. At fourteen months' follow-up, imaging demonstrated marked improvement with reconstitution of normal bony anatomy. This case illustrates that CAPNON can occur in infancy, may behave more aggressively than in adults, and can present with an atypical soft macroscopic consistency. Awareness of this entity is essential to guide appropriate surgical planning and postoperative management in the paediatric population.

Introduction

Calcifying pseudoneoplasms of the neuraxis (CAPNON) are rare, non-neoplastic calcified lesions of the central nervous system, thought to represent a reactive or inflammatory process rather than a true neoplasm [1, 2]. First described by Rhodes and Davis in 1978 [3], fewer than 150 cases have been reported in the literature. Due to their rarity and nonspecific imaging characteristics, CAPNON lesions are frequently misdiagnosed as more common intracranial or spinal tumours, including meningioma, chondrosarcoma, and chordoma [4].

The exact aetiology remains unclear. CAPNON is hypothesised to arise as a reactive response to trauma, inflammation, infection, or adjacent neoplasia, supported by its benign behaviour and characteristic fibro-osseous histology [5]. These lesions are typically slow-growing and densely calcified, most commonly occurring in the supratentorial region, though they may arise anywhere along the neuraxis [4]. Male predominance has been reported [1, 2, 6–11].

Paediatric cases are exceedingly uncommon. To date, only few prior paediatric case has been reported, and no cases in infancy have previously been described [6]. We present the youngest reported patient with CAPNON involving the craniocervical junction, highlighting the clinical, radiological, and histopathological features, the atypical intraoperative macroscopic findings, and the postoperative management of craniocervical instability.

Case Presentation

A male infant born extremely prematurely at 27 + 6 weeks' gestation required prolonged neonatal intensive care. His neonatal course was complicated by chronic lung disease of prematurity, grade I intraventricular haemorrhage, and progressive ventriculomegaly.

At five months of age, he suffered an out-of-hospital cardiac arrest with an estimated downtime of 20 minutes, resulting in hypoxic–ischaemic brain injury, likely secondary to progressive hydrocephalus. Initial neurosurgical management included insertion of an external ventricular drain, followed by ventriculoperitoneal (VP) shunt placement at six months of age.

Subsequent neuroimaging at six months identified a lesion at the craniocervical junction. Serial CT imaging at nine months demonstrated progressive heterotopic calcification extending from C1 to C3, most pronounced at C1–C2, with increasing density and midline extension (Fig. 1A, B). The spinal canal diameter was reduced to approximately 6 mm at C1. The left C2 lamina was completely replaced by calcified tissue, and an enlarging anterolateral mass was noted at C2, with no extension below C3. MRI confirmed severe spinal cord compression and near-complete obliteration of cerebrospinal fluid spaces at this level (Fig. 2A, B). By eight months of age, the patient had developed cervical myelopathy, respiratory compromise with recurrent apnoeic episodes, and unsafe swallowing.

(A, B) Preoperative images show a heterogeneous low-signal lesion in the left parapharyngeal region and severe canal stenosis at C2 with marked spinal cord compression and near-complete obliteration of CSF spaces. (C, D) One-month postoperative images demonstrate reduced calcific burden, particularly on the left, with persistent partial canal stenosis and cord compression. (E, F) Nineteen-month postoperative images show marked interval improvement, with significant resolution of canal stenosis and reduction of the extra-axial calcified lesion.

Based on the CT appearance of dense calcification, the lesion was expected to require drilling for removal, and C1–C3 laminectomy with tumour excision was planned. However, intraoperatively, the arches of C1 and C2 were found to have completely liquefied; the lesion consisted of thick, opaque, bright-white fluid rather than solid calcified material. Partial debulking was performed at 9 months of age, achieving significant decompression despite residual tissue. Because both vertebral arches lacked solid osseous support, the findings were considered with C1–C2 instability.

Histopathological examination of the resected material confirmed CAPNON, demonstrating extensive dystrophic calcification, osseous metaplasia, and a foreign-body giant cell reaction, with no evidence of neoplasia (Fig. 3).

Postoperatively, given the young age of the patient and the absence of rigid osseous support, definitive craniocervical fusion was deferred. The child was managed with prolonged rigid cervical immobilisation. Following fourteen months in a cervical collar, repeat imaging demonstrated marked improvement: significant reduction in lesion size, reconstitution of normal bony anatomy, and stable spinal canal dimensions without evidence of instability (Fig. 1E, F; Fig. 2E, F). The patient remains under close neurosurgical and orthopaedic surveillance. Surgical fusion has not been required.

Discussion

This case represents the youngest reported patient with CAPNON and the first documented case involving the craniocervical junction in an infant. Paediatric cases of CAPNON are extremely rare; to our knowledge, only few prior paediatric case has been published [6], making this case clinically and pathologically noteworthy.

CAPNON can occur anywhere along the neuraxis. Clinical presentation depends on lesion location and size, with symptoms resulting from mass effect. Intracranial lesions most commonly present with seizures or headache, and spinal lesions with pain or myelopathy [6, 8–11]. Our patient presented atypically with apnoea, cardiorespiratory compromise, and myelopathy secondary to severe craniocervical stenosis, reflecting the critical location of the lesion.

In adults, CAPNON is generally slow-growing, and conservative management may be appropriate for asymptomatic cases [6, 17]. In contrast, our patient demonstrated rapid lesion progression, likely reflecting active bone remodelling associated with skeletal immaturity. This behaviour parallels observations in other ossified lesions in young children and underscores the importance of close monitoring and low threshold for surgical intervention in paediatric patients.

Radiologically, CAPNON typically appears as a densely calcified mass on CT, with peripheral calcification and minimal bone erosion; MRI typically shows hypointensity on T1- and T2-weighted sequences with variable enhancement [6, 8, 11–16]. Our case demonstrated these typical imaging features. However, a critical finding was the complete discordance between the preoperative imaging appearance and the intraoperative macroscopic consistency: the lesion, which appeared densely calcified on CT, consisted entirely of thick, opaque, bright-white liquid in consistency at surgery. This has not been described in prior CAPNON reports and carries important surgical planning implications – surgeons should be prepared for this possibility when operating on CAPNON at the craniocervical junction in infants.

Histopathologically, CAPNON characteristically shows calcified fibro-osseous tissue with psammomatous or dystrophic calcification, chondromyxoid matrix, and foreign-body giant cell reaction, without mitotic activity or necrosis [1, 3, 12–16]. Our specimen demonstrated these typical features, confirming the diagnosis despite the atypical macroscopic appearance.

Differential diagnoses for calcified craniocervical lesions include meningioma, chordoma, chondrosarcoma, schwannoma, vascular malformations, granulomatous infections, and other calcified lesions [1, 17]. Histopathological examination remains essential to distinguish CAPNON from these entities.

Surgical resection is the mainstay of treatment for symptomatic CAPNON, providing both decompression and definitive histological diagnosis. While complete resection is generally preferred, the literature – including a systematic review by Stienen et al. – suggests that incomplete resection does not significantly increase recurrence risk [6]. Our case supports this observation, with marked radiological regression of residual lesion at fourteen months. This suggests that partial resection with close follow-up may be a safe and effective strategy when complete excision is not feasible.

Postoperative craniocervical instability is an uncommon but important complication following decompressive surgery at the craniocervical junction, particularly in the context of pre-existing osseous disruption. In our patient, the complete liquefaction of the C1–C2 arches rendered the spine inherently unstable following surgery. Definitive fusion was deferred given the patient's young age and small size, and prolonged rigid cervical immobilisation was selected as a temporising strategy. The spontaneous reconstitution of bony anatomy at follow-up imaging, without requirement for instrumented fusion, represents a favourable outcome not previously described in this context and may reflect the significant remodelling capacity of the infantile skeleton.

Conclusion

CAPNON should be considered in the differential diagnosis of calcified craniocervical lesions in infants, despite its rarity. This case illustrates that paediatric CAPNON may behave more aggressively than in adults and can present with atypical macroscopic findings, including a soft, liquid-like consistency despite a densely calcified CT appearance. Surgical decompression is essential for symptomatic lesions, and partial resection may be sufficient with close follow-up. Craniocervical instability is a recognised postoperative risk; in young infants, prolonged rigid immobilisation may allow spontaneous skeletal remodelling, potentially avoiding the need for definitive surgical fusion.

Declarations

Ethics Approval and Consent to Publish

Written informed consent for publication was obtained from the patient's parent/legal guardian. All procedures were conducted in accordance with the ethical standards of Alder Hey Children's Hospital NHS Foundation Trust and with the 1964 Helsinki Declaration and its later amendments. Formal ethical committee review was not required for this single case report under NHS Health Research Authority criteria.

Informed Consent

Written informed consent for publication of this case report, including clinical details and imaging, was obtained from the patient's parent/legal guardian.

Competing Interests

The authors declare no competing interests.

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Data Availability

The data supporting the findings of this case report are available from the corresponding author upon reasonable request, subject to applicable NHS governance requirements and patient confidentiality.

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Author Contribution

Authors' Contributions YK contributed: data collection, literature review, drafting and revision of the manuscript. LDL, MM & MM contributed equally to images data collection & analysis, and critical revision manuscript. FR contributed neuropathological expertise, histopathology pictures and critical revision of the manuscript. CP contributed to clinical management, supervision, and critical revision. All authors approved the final manuscript.

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Figures

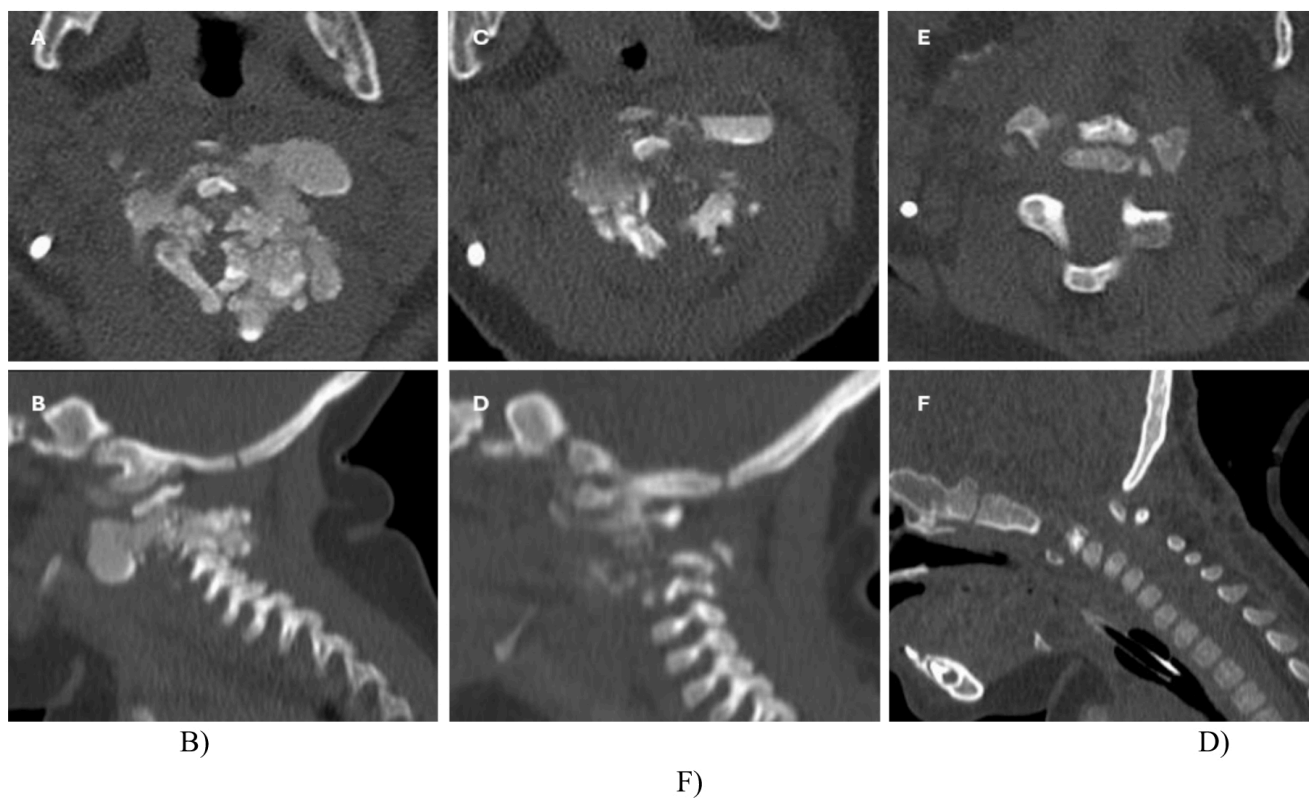


Figure 1

CT imaging demonstrating interval changes in calcified lesion at the craniovertebral junction.

(A, B) Preoperative axial and sagittal images show extensive heterotopic calcification from C1–C3 with severe spinal canal stenosis. (C, D) One-month postoperative images demonstrate partial debulking of the lesion with partial canal stenosis and a residual anterior component exhibiting a milk of calcium cyst raising the possibility of tumoural calcinosis. (E, F)

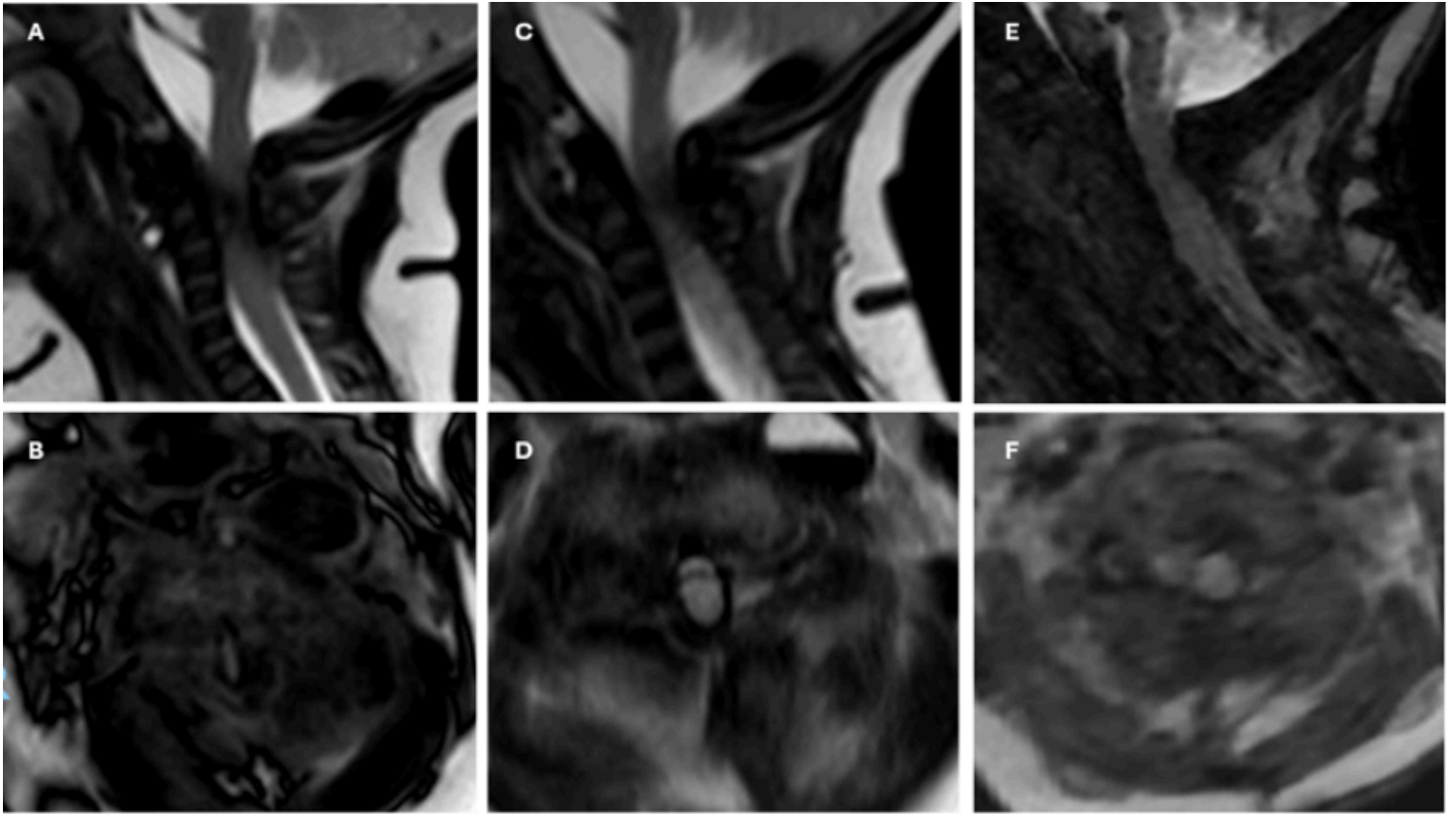


Figure 2

MRI sequences demonstrating interval changes in the craniovertebral junction lesion and spinal canal compromise.

(A, B) Preoperative images show a heterogeneous low-signal lesion in the left parapharyngeal region and severe canal stenosis at C2 with marked spinal cord compression and near-complete obliteration of CSF spaces. (C, D) One-month postoperative images demonstrate reduced calcific burden, particularly on the left, with persistent partial canal stenosis and cord compression. (E, F) Nineteen-month postoperative images show marked interval improvement, with significant resolution of canal stenosis and reduction of the extra-axial calcified lesion.

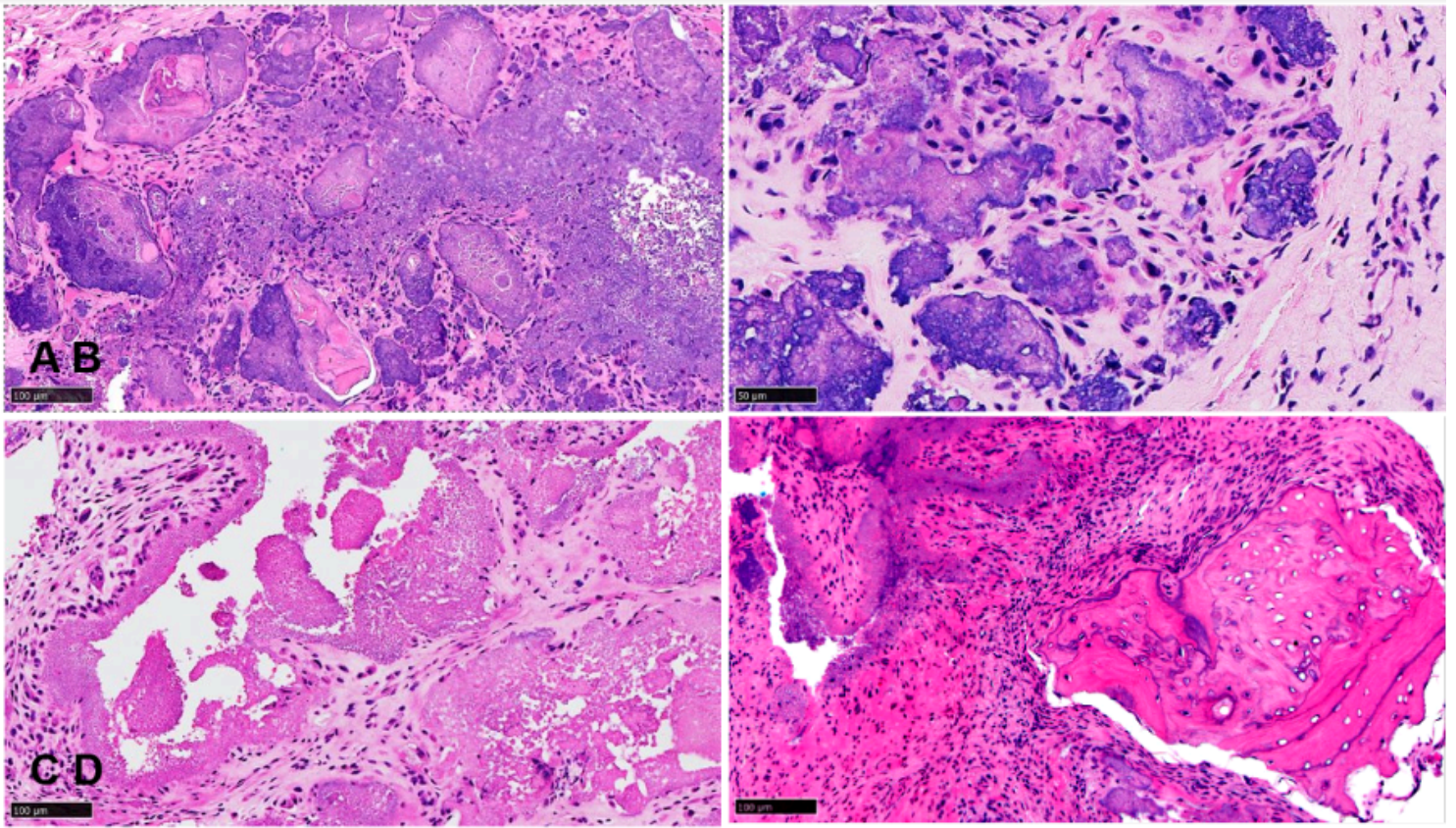


Figure 3

Histopathological images. The lesion is composed of fibrous connective tissue with dystrophic calcification, granular calcium deposits, and metaplastic bone (A, haematoxylin–eosin, $\times 10$; B, haematoxylin–eosin, $\times 20$). Granular calcium deposits delineating cystic spaces (C, haematoxylin–eosin, $\times 20$). Devitalised metaplastic bone is also present (D, haematoxylin–eosin, $\times 20$). These features are diagnostic of CAPNON.