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(CASE REPORT)

Uncommon presentation: Extra-axial medulloblastoma in the cerebellopontine angle in an adult: Case report and literature overview

Meriem Kajeou *, Yasser Arkha

Department of Neurosurgery, Faculty of Medicine and Pharmacy of Rabat, Specialty hospital in Rabat, Mohammed V University in Rabat, Rabat, Morocco

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Abstract

Introduction: The medulloblastoma is a pediatric malignant primitive neuroectodermal tumor. It's occure rarely in adults, accounting for less than 1% of brain tumors. The typical location is the cerebellar vermis toward the fourth ventricle. The extra-axial location of the cerebellopontine angle is extremely rare.

Case report: Reporting a case of extra-axial medulloblastoma at the cerebellopontine angle in a 23-year-old right-handed adult with no significant medical history. The patient presented with gradually worsening headaches, dizziness, and gait disturbances. Clinical examination revealed a conscious patient with a GCS of 15, exhibiting a vestibular cerebellar syndrome, right hemiparesis 4/5, grade II right facial paralysis, right-sided deafness, and bilateral grade I papilledema.

The patient underwent initial surgery, including a primary ventriculoperitoneal shunt to address hypertension syndrome, followed by a retro-mastoid approach to the cerebellopontine angle with macroscopically complete tumor resection. The postoperative course was uneventful, and the histopathological diagnosis confirmed medulloblastoma of the cerebellopontine angle. The patient was subsequently referred for oncology.

Conclusion: The extra-axial medulloblastoma at the cerebellopontine angle is an extremely rare tumor. It should be considered in the differential diagnosis of tumors involving the cerebellopontine angle.

Keywords: Cerebellopontine angle; Médulloblastoma; Rare; Surgery

1. Introduction

Medulloblastoma is the most common malignant tumor of the brain in the pediatric population. It is a solid, homogenously enhancing, midline tumor of the posterior fossa [1]. In the existing literature, only a limited number of cases involving cerebellopontine (CP)-angle medulloblastoma have been reported. The extra-axial site of this tumor remains a rarity [2, 3]. In adults, the CP angle can host various tumors, with the most prevalent being schwannomas, meningiomas, and epidermoid cyst tumors, followed by metastatic lesions. The occurrence of medulloblastoma at the CP angle is notably rare, and the presence of a solely extra-axial CP angle medulloblastoma in an adult, as documented in this paper, is extremely uncommon [4]. Studies published on adult medulloblastoma are typically retrospective due to its infrequency in adult populations, and they predominantly focus on midline occurrences, specifically the cerebellar vermis [5, 6].

* Corresponding author: Kajeou meriem

We present an exceptionally rare case of medulloblastoma in the adult CPA. This case emphasizes the presence of an extremely rare extra-axial medulloblastoma in this unconventional location, thereby complicating its diagnosis. Additionally, it highlights the challenges encountered when relying on imaging for medulloblastoma diagnosis, especially in adults.

2. Case report

We report the case of extra-axial medulloblastoma at the CP angle in a 23-year-old right-handed adult with no significant medical history. The patient presented with gradually worsening headaches, dizziness, and gait disturbances over a duration of two months. Clinical examination revealed a conscious patient with a GCS of 15, exhibiting a vestibular cerebellar syndrome, right hemiparesis 4/5, grade II right facial paralysis, right-sided deafness, and bilateral grade I papilledema.

The radiological exploration favored an extra-axial lesion process in the right CP angle with a seems like broad-based attachment, showing isosignal on T2 and Flair images, hyposignal on T1 images, diffusion hyperintensity, and mild enhancement after Gadolinium injection. This process induces a significant mass effect on the pons and the fourth ventricle, pushing it towards the contralateral side, with moderate upstream ventricular dilatation. It produces a mass effect on the cerebellar parenchyma without perilesional edema, and it comes into contact with the patent basilar artery and the ipsilateral transverse sinus, which remains patent. Based on the anatomical and radiological findings, a provisional diagnostic hypothesis of meningioma, was considered**.** [Figure 1].

The patient underwent surgery in the Park-bench position, with a standard right retro-sigmoid approach. A right retrosigmoid suboccipital craniotomy with complete excision of the lesion was performed. Intraoperatively, the tumor appeared grayish, solid but soft and friable, and easily suctionable **[**Figure 2]. There was a clear cleavage plane between the tumor and the cerebellum. The frozen section favored an undifferentiated round cell tumor. Progression in tumor excision was achieved using bipolar cautery, gentle suction, and an ultrasonic aspirator. The tumor exhibited some hemorrhagic areas with multiple coagulated intra-tumoral vessels. During the procedure, visualization of the vein of Dandy, nerves V, VI, VII, mixed nerves, the superior cerebellar artery, and the basilar trunk was accomplished. Careful excision was performed around the acoustic-vestibulo-facial bundle, as well as around the mixed nerves inferiorly and nerve V superiorly. Macroscopic excision was deemed complete [Figure3]. His postoperative period proceeded without any incidents. There was no neurological deterioration, and his cerebellar signs also improved gradually**.** The postoperative CT scan showed no residual mass or postoperative complications except a small hyperdensity in CP angle [Figure 4].

The histopathological assessment revealed a hypercellular tumor with round cell morphology and frequent mitosis. Immunohistochemical study demonstrated positivity for synaptophysin and TP53 mutation positivity. Imprint smears displayed small clusters and scattered pleomorphic tumor cells against a desmoplastic stroma [HE×10 and HEx5]. These findings are indicative of a classic medulloblastoma, classified as grade IV according to the 2021 WHO classification [Figure 5].

Considering that the histopathological diagnosis contradicted our preoperative impression, the specimens underwent a reevaluation by a second pathologist, but the diagnosis remained unchanged. Following the communication of the histopathological results, a spinal cord MRI was conducted, revealing no evidence of spinal cord metastasis. Subsequently, the patient was referred for additional oncological care, including radiotherapy and chemotherapy, and will undergo regular follow-up.

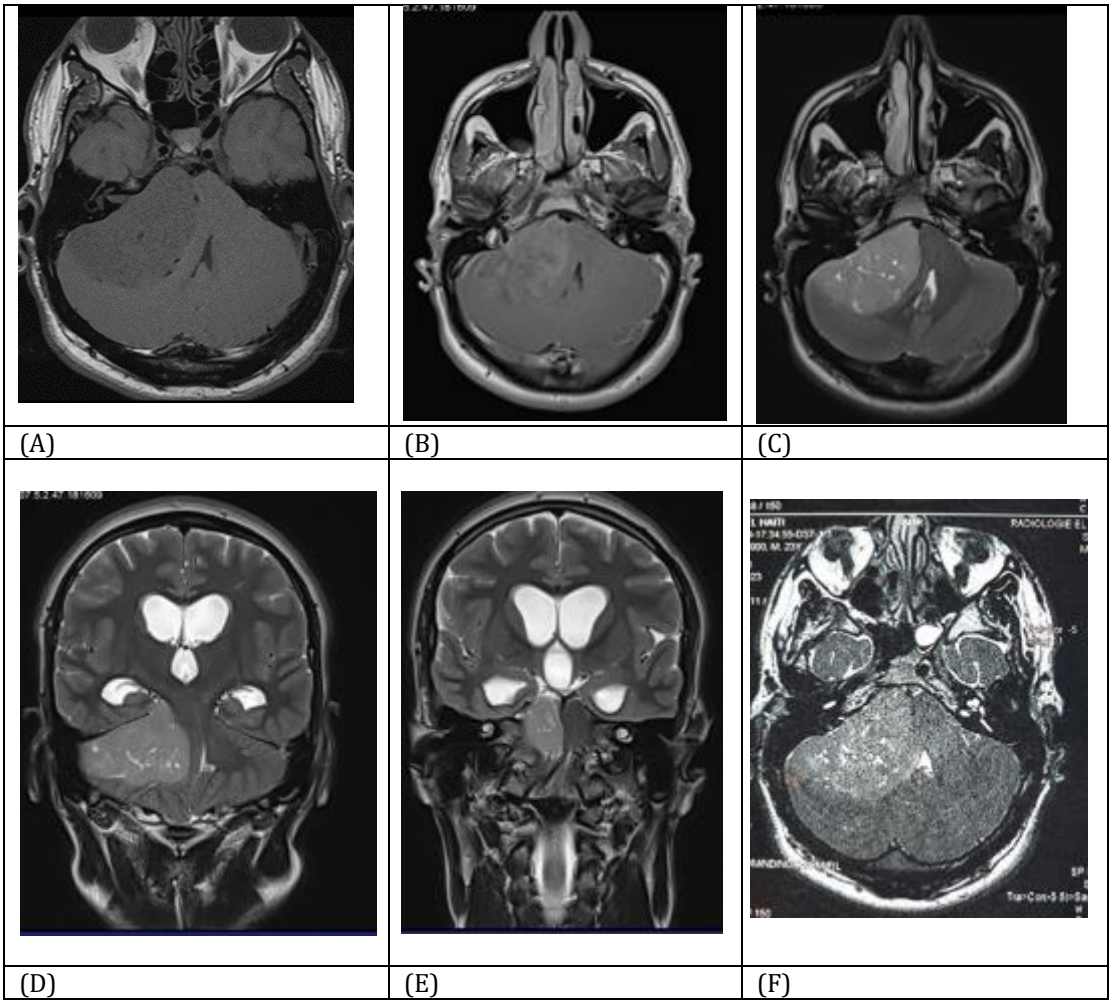


Figure 1 The preoperative MRI images show : An isosignal on T2 and FLAIR images (C, D, E,F), hyposignal on T1 image (A), and mild enhancement after Gadolinium injection (B)

Figure 2 Intraoperative image showing the tumor's appearance with a cleavage plane (yellow arrow)

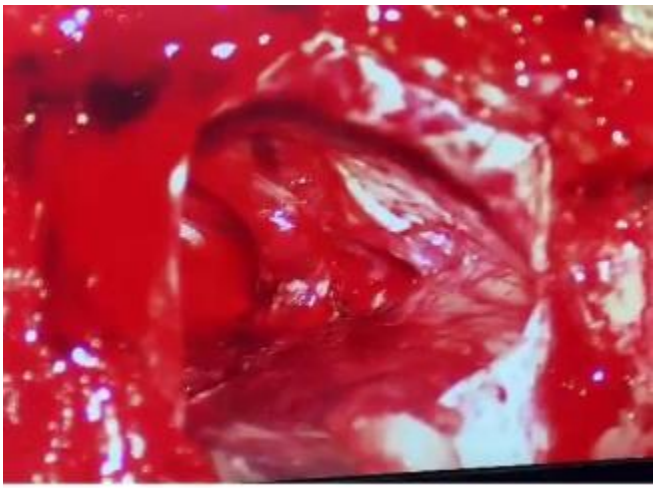


Figure 3 Intraoperative image illustrating the excision cavity with complete removal of the lesion

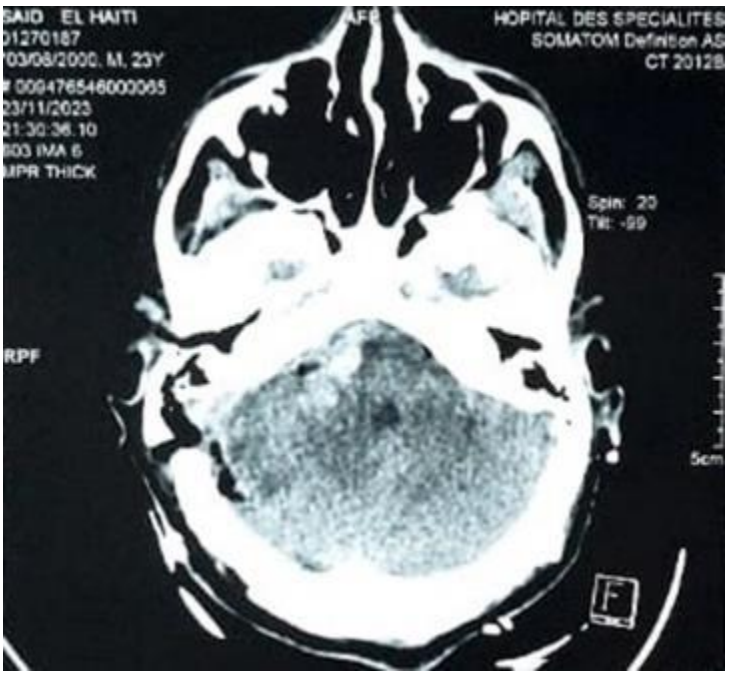
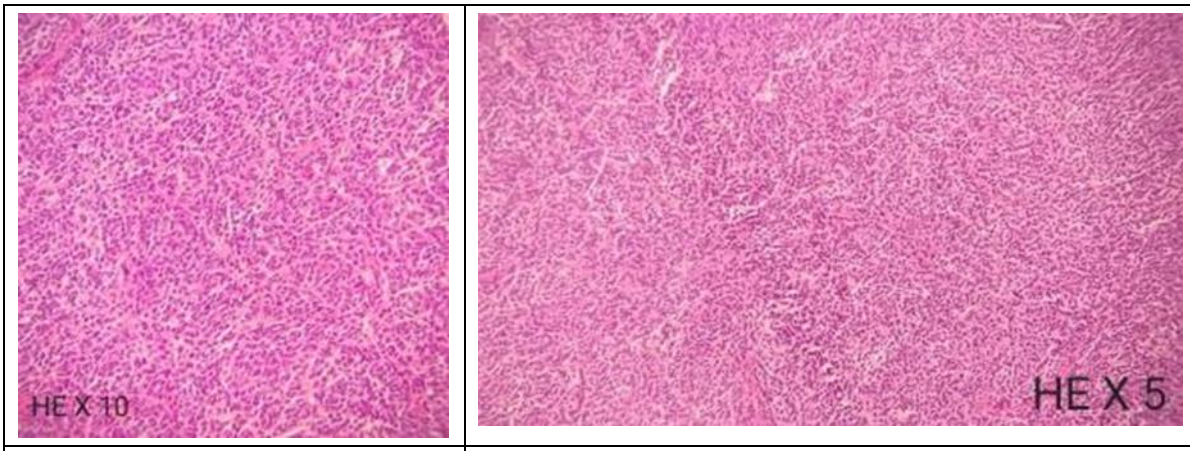
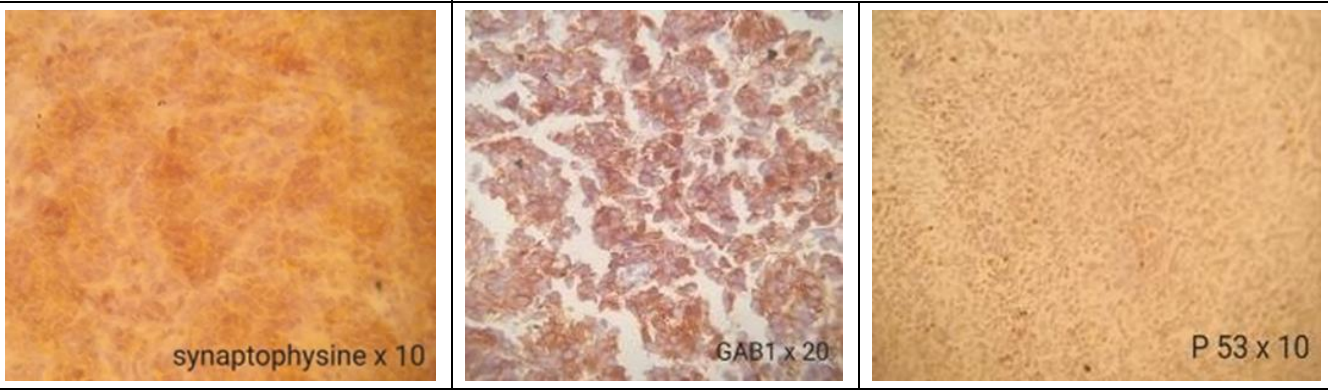


Figure 4 Postoperative head computed tomography scan, axial view, showing complete resection of the tumor



(A) (B)



(C) (D) (E)

Figure 5 Imprint smears displayed small clusters and scattered pleomorphic tumor cells against a desmoplastic stroma (A,B), Synaptophysin is positive (C), Anti-GAB1 antibody (Clone BSB-155, BioSB) (D), Negative for anti-P53 antibody (Clone DO7, BioSB) (E), These findings are indicative of a classic medulloblastoma, classified as grade IV according to the 2021 WHO classification

3. Discussion

Medulloblastoma is predominantly a pediatric tumor that frequently arises in the cerebellar vermis. Common lesions in the cerebellopontine angle typically include acoustic neuromas, followed by meningiomas, primary cholesteatomas, and epidermoid tumors. Together, these lesions account for up to 98% of tumors in this region [7, 8]. However, a purely extra-axial location of medulloblastoma at CP angle other than these usual locations in an adult patient is quite exceptional [9]. Distinguishing these diagnoses can be difficult, due to the absence of clear clinical and radiological features. Moreover, when a patient with cerebellopontine medulloblastoma experiences deafness, as evidenced in this case, the differentiation from acoustic schwannoma becomes even more complex [10]. The extra-axial location of the tumor in the cerebellopontine angle in adults makes our patient's case quite rare.

The origin of medulloblastomas in the CP angle remains unclear. CP angle medulloblastomas have been proposed to arise from residue in the external granular layer of the cerebellar hemisphere, specifically the flocculus facing the cerebellopontine. They can also emerge from the proliferation of the lateral medullary velum germinal residues, then protruding into the cerebellopontine. Medulloblastomas found in the fourth ventricle are often assumed to develop laterally to the cerebellopontine through the Luschka foramen or direct exophytic growth from the origin site at the surface of the cerebellum or pons [11,12].

The radiological features of medulloblastoma are typical: it exhibits an iso- or hypointense signal on T1-weighted images, is heterogeneous on T2-weighted images, and demonstrates homogeneous enhancement after gadolinium administration, sometimes accompanied by a central hemorrhagic zone [13].

In our case, the preoperative diagnosis of cerebellopontine angle meningioma was influenced by the patient's age at presentation and the imaging characteristics of the lesion, which was well-defined, broad-based, extra-axial, and exhibited homogeneous enhancement. Medulloblastoma in adults is a rare entity, accounting for only 1% of primary brain tumors in adults. while medulloblastomas, most often desmoplastic, are usually found in the cerebellar hemispheres, in adults they are rarely found in the cerebellopontine angle (JE DOIS TROUVER UNE REFERENCE).To date, only 10 cases of extra-axial medulloblastoma have been reported in the adult literature [13].

The table [Table 1] describes previously reported CP angle medulloblastomas, including their presentation, management, and outcome, in comparison to the current case.

Table 1 A review of the literature on extra-axial cerebellopontine angle medulloblastomas.

Author	Year	Age/Sex	Clinical features	Radiological diagnosis	Histopathology	Treatment
Becker RL et al. [14]	1995	Case 1 : 32/female Case 2 : 52/female	Case 1 : Hearing loss, ataxia, diplopia, nausea Case 2 : NA	Case 1: Meningioma/ Schwannoma Case 2 : Meningioma/ schwannoma	Medulloblastoma Medulloblastoma	NA NA
Akay KM et al. [14]	2003	21/male	Headache, Vomiting, Ataxia, Bilateral papilledema, Hemiparesis, Hemianesthesia	Impaired hearing on audiometry Hypointense on T1-weighted images and hyperintense on T2-weighted images	Medulloblastoma	Left suboccipital craniectomy in the prone position. The lesion was exposed through the left CP angle. Partial excision Radiotherapy and Chemotherapy
Salu Gi JG et al. [14]	2004	40/male	Headache, vomiting, hearing difficulties	Meningioma	Desmoplastic medulloblastoma	NA
Fallah A et al. [14]	2009	47/male	Headache, nausea/ vomiting	Meningioma	Medulloblastoma	Total excision Radiotherapy
Furtado SV et al. [14]	2009	32/male	Headache, vomiting, gait unsteadiness, papilledema, dymetria,dysdiadokinesia, ataxia	Meningioma	Medulloblastoma classic	Total excision Adjuvant therapy
Singh M et al. [14]	2011	22/male	Headache, vomiting, facial weakness, papilledema, ataxia	Extra axial Mass	Medulloblastoma classic	Total excision
Bahrami E et al. [14, 15]	2013	23/male	Hearing loss, Nausea and Vomiting, Ataxia	Hypointense on T1-weighted images and hyperintense	Desmoplastic medulloblastoma	Retromastoid craniectomy, and Total resection,

				on T2-weighted images, The lesion enhanced on T1 weighted MRI images after injection of gadolinium		and Radiotherapy
Spina et al. [16]	2013	Case A: 22 M Case B: 26 F	Case A: Headache, ataxia, hearing loss in her left ear, dizziness, and tinnitus as of the last 3 months Weakness of the right arm, slight Left nystagmus and a mild peripheral deficiency of the left VII-CN nerve. Case B: Chronic headache	Case A: Hyperin - tense T2-weighted images, hypointense T1-weighted images Case B: Hypointense in T1-weighted images and isointense in T2- weighted images, Edema in the anterior superior right cerebellar hemisphere	Classic extra-axial MB in the CPA Case A: Classic type with nodular aspects Case B: Classic type of MB infiltrating the arachnoid	Case A: retrosigmoid craniectomy and gross total tumor resection and radiotherapy. Case B: right retrosigmoid craniectomy with gross-total resection. Radiation therapy
Goudihalli SR et al. [14]	2018	50/male	Facial asymmetry, hearing loss	Schwannoma	Medulloblastoma classic	Total excision Follow up : vegetative
Ninh Ba Doan et al. [17]	2018	29/male	Syncopal episode and headache for the last 2 weeks	Normal neurological exam MRI: Tentorial dural-tail sign	Extra-axial midline tentorial MB grade IV SHH type Desmoplastic	Unremarkable postoperative period Radiation and adjuvant chemotherapies (vincristine, cisplatin, and cyclophosphamide).
Ratha V et al. [14]	2019	42/female	Headache, ataxia	Heterogeneous lesion exhibiting hypo-intensity compared to gray matter on T1-weighted images and hyperintensity on T2-weighted images, with heterogeneous enhancement of the lesion after contrast administration	Medulloblastoma classic	Total excision and radiotherapy
Pant Ishita (18)	2020	30/female	Headache, nausea/ vomiting	MRI f the brain revealed a large extra-axial mass in the right CP	Desmoplastic Medulloblastoma	Total excision Radiotherapy

				angle with a broad dural base causing mass effect on the underlying cerebellum and brain stem with resultant compression and displacement of fourth ventricle causing upstream obstructive hydrocephalus. The mass showed moderate heterogeneous contrast enhancement with prominent dural enhancement Diffusion-weighted images showed restricted diffusion suggesting dense cellularity		
Present case, 2023	2023	23/male	Headache, Nausea and Vomiting, Ataxia, Bilateral papilledema, Hemiparesis.	Isosignal on T2 and Flair images, hyposignal on T1 images, diffusion hyperintensity, and mild enhancement after Gadolinium injection	Grayish lesion, solid but soft and friable, and easily suctionable.There was a clear cleavage plane between the tumor and the cerebellum. The histopathological result favored a classic medulloblastoma, Grade IV according to the WHO 2021 classification.	Right retrosigmoid approach. Total excision, No complication. Oncological management

Medulloblastomas are known to metastasize through the cerebrospinal fluid to the spinal canal, leptomeninges, and supratentorial regions. The metastasis rate in medulloblastomas varies between 38% and 60% in various series, with the spinal canal being the most common site at approximately 58% (3; 19). However, spinal metastasis from cerebellopontine angle medulloblastoma has not been reported till now. In our case, no metastasis was detected in the cerebrospinal fluid (CSF) or at the spinal level.

Radical surgery is the primary therapeutic approach for cerebellopontine angle medulloblastoma. The combination of complete gross resection with radiotherapy has demonstrated its efficacy in improving long-term prognosis. The 5-year survival, just 30% in 1960s, has nearly doubled, owing to more innovative surgical techniques and effective adjuvant chemotherapies and radiotherapies (20,21). In our case, the patient underwent macroscopically complete resection followed by radiotherapy and exhibited a favorable outcome.

4. Conclusion

Over the years, medulloblastomas have exhibited significant variability in terms of presentation, radiological diagnosis, and biological behavior. Although generally regarded as a common pediatric intra-axial tumor, the increasing isolated reports of its extra-axial presentation in the CP angle region necessitate its inclusion in the differential diagnosis of CPA tumors. This exceptionally rare occurrence in adults at this unusual location can lead to confusion in the accurate diagnosis and delay the initiation of necessary adjuvant therapy

Finally, the extra-axial site of this tumor is extremely rare but must be considered in the differential diagnosis of extra-axial CP angle lesions.

Compliance with ethical standards

Disclosure of conflict of interest

No conflict of interest to be disclosed.

Statement of informed consent

Informed consent was obtained from all individual participants included in the study.

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