Radiology Case Reports 16 (2021) 2509–2513

Available online at www.sciencedirect.com

ScienceDirect

journal homepage: www.elsevier.com/locate/radcr

Case report

A case of lipoma, calcification, and brain malformations in the midline of the skull

Hideto Kanazawa, MD[a,b,*], *Kenji Gonda, MD, PhD*[b,c,d], *Takamitsu Tamura, MD, PhD*[a,b], *Seiichi Takenoshita, MD, PhD*[d]

[a] *Department of Neurology, Daido Central Hospital, 1-1-37 Asato, Naha, Okinawa 902-0067, Japan*
[b] *Daido Obesity and Metabolism Research Center, Naha, Okinawa, Japan*
[c] *Department of Gastrointestinal Tract Surgery, Fukushima Medical University, Fukushima, Japan*
[d] *President of Fukushima Medical University, Fukushima, Japan*

ARTICLE INFO

Article history:
Received 11 March 2021
Revised 26 May 2021
Accepted 28 May 2021

Keywords:
Cranial neural crest cells
Malformation
Symmetry
Pericallosal lipoma

ABSTRACT

Few cases of pericallosal lipoma with several other lesions, including specific forms of calcification and brain malformations, have been reported. We present the case of an asymptomatic 83-year-old man with a pericallosallipoma with peculiar symmetrical morphology in the midline of the skull. We posit that the lesions began forming in the very early embryonic period and were closely associated with the cranial neural crest cells. We report the neuroradiological findings of this characteristic lesion and discuss several literature reviews on the process of its formation.

Introduction

The pericallosal lipoma (PCL) is extremely rare, comprising 0.2%-0.5% of all brain tumors [3,12]. Lipomas are often malformed because they rarely infiltrate surrounding tissues. In addition to the PCL, we identified various lesions in our patient such as excess adipose tissue in the arachnoid, calcification, and cingulate gyrus with partial loss bilaterally. Furthermore, these parts were displayed remarkable symmetry. We present the neuroradiological findings of this rare case and discuss the available literature.

Case report

An 83-year-old male patient complained of low back pain subsequent to a fall and presented to our hospital's orthopedics department. A non-contrast-enhanced computed tomography (CT) scan of the head was conducted to exclude any acute intracranial abnormality; while there were no acute lesions, abnormalities were incidentally identified in the midline of the skull. The CT revealed a low-density median mass with a central density of -60 to -120 HU. The mass was located symmetrically between the lateral ventricles with respect to

* Corresponding author.
E-mail addresses: opirineppu@gmail.com (H. Kanazawa), gondake@fmu.ac.jp (K. Gonda), takamitsutamura0926@gmail.com (T. Tamura), akiko-t@fmu.ac.jp (S. Takenoshita).
https://doi.org/10.1016/j.radcr.2021.05.073

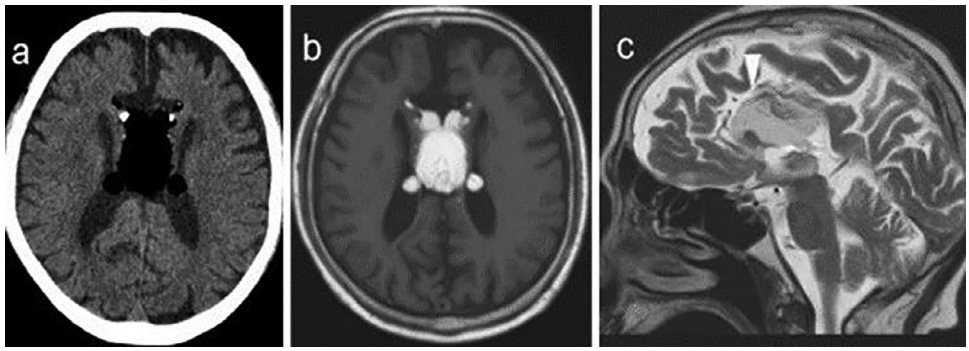

Fig. 1 – Axial non-enhanced computed tomography (A) revealed a low-density median mass midway between the lateral ventricles. Axial (B) unenhanced T1-weighted and midline sagittal (C) T2-weighted images revealed a high-signal area (white arrow) between the lateral ventricles.

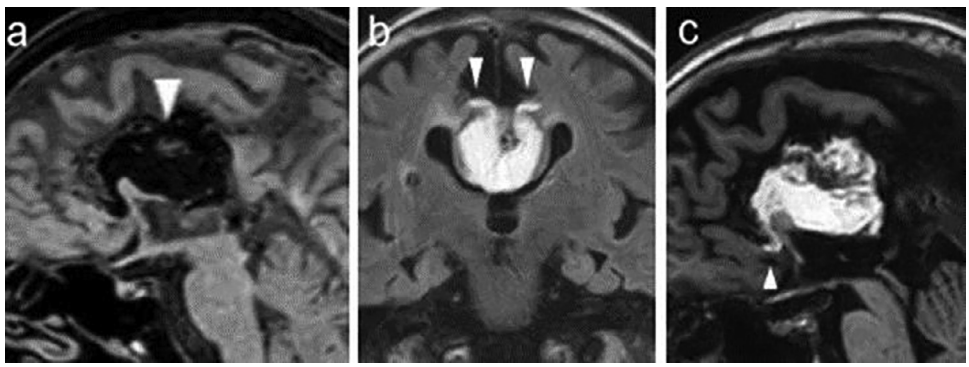

Fig. 2 – Midline sagittal (A) fat-saturated T1-weighted images eliciated no signal (white arrow). A coronal (B) unenhanced T1-weighted image revealed adipose tissue invading the cingulate gyrus (white arrow). Similar findings were observed (C) at the paraterminal gyrus (white arrow).

the skull midline and extended to the choroid plexus (Fig. 1A). Calcification was observed bilaterally on the surface of the mass. The patient was referred to the department of neurology. He was awake, alert, and oriented, and his neurological exam was normal. He also had no relevant neurological history. Radiological evaluation, including magnetic resonance imaging (MRI), was conducted to examine the lesion. The head MRI revealed a high-intensity, non-enhanced median mass that continued from the lateral ventricle to the choroid plexus on the T1 sequences. The size was 55 mm in the anterior-posterior direction, 35 mm in the coronal plane, and 45 mm in the cranio-caudal direction (Fig. 1B). On the sagittal T2 image, the lesion was located on the dorsal side of the corpus callosum which was formed by only the rostrum (Fig. 1C).

The intrinsic T1 hyperintense signal was completely suppressed on the fat-saturated sequences, consistent with adipose tissue (Fig. 2A). A coronal unenhanced T1 weighted MRI image revealed that the adipose tissue invaded the arachnoid mater from the cingulate gyrus to the frontal region in a symmetrical sheetlike form (Fig. 2B). Similar findings were observed at the paraterminal gyrus on sagittal unenhanced T1-weighted images (Fig. 2C).

To observe the peculiarly shaped calcified part in more detail, we created a 3D volume-rendered reconstructed image. The calcification was symmetrical with respect to the midline and was divided into left and right sides. They were hook-shaped from the superior aspect of the lipoma to the paraterminal gyrus (Figs. 3A and B). We have included imaging from cases of similar calcification associated with intracranial lipoma for reference [10] (Fig. 3C).

The cerebrovascular system was observed using non-contrast MR angiography. To understand the positional relationship with the adipose tissue and calcification better, the adipose tissue imaged by MRI and the calcification imaged by CT were reconstructed in 3D (Fig. 4). In this image, the adipose tissue is shown in yellow, the calcification in white, and the vascular system in red.

Following the left and right internal carotid arteries, the anterior cerebral artery branches normally, subsequently ascending along the anterior surface of the adipose mass while curving backward, with the bilateral pericallosal and callosomarginal arteries branching normally (Fig. 4). Each of these vessels passed through the medial and lateral dorsal sides of a large central oval adipose mass, releasing several small vessels towards the mass and finally toward the frontal and parietal lobes. No details could be observed inside the mass.

Two brain malformations were also observed. On sagittal fat-saturated T1-weighted MRI images, hypoplasia of the

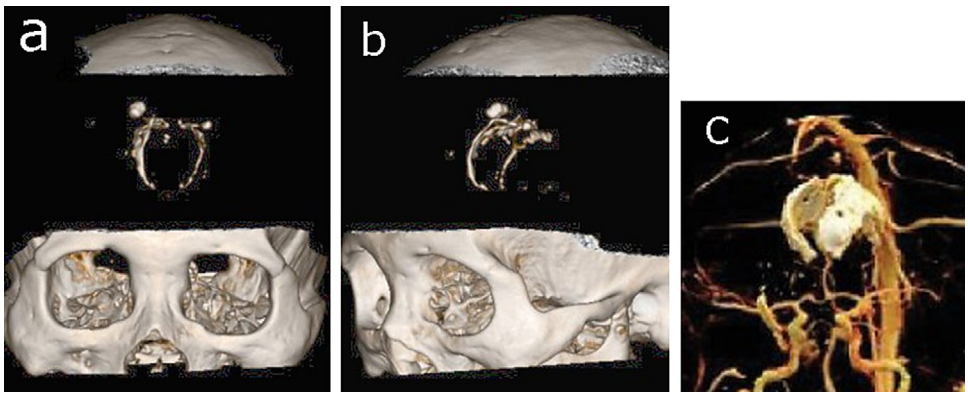

Fig. 3 – Coronal (A) and oblique (B) 3D computed tomography volume-rendered reconstructed image. Curvilinear calcification was present around the adipose mass. Similar calcification was reported in a previous case in the literature (C) [10].

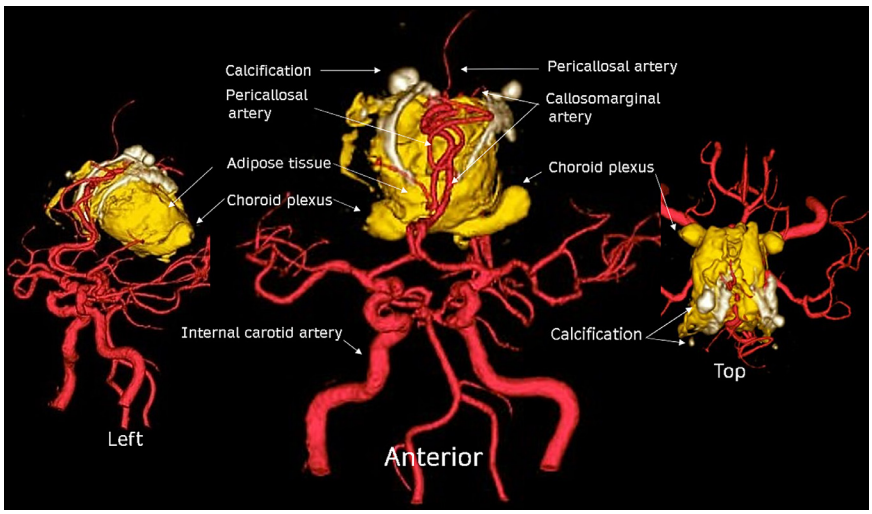

Fig. 4 – The adipose tissue (yellow), imaged by magnetic resonance, and the calcification (white) imaged by computed tomography, were reconstructed in 3 dimension and synthesized with the vascular (red) image(color version of figure is available online).

corpus callosum which only formed the rostrum was observed (Fig. 5A). The anterior ventral cingulate gyrus and the anterior dorsal cingulate gyrus were similarly missing bilaterally (Figs. 5B and C).

Discussion

The mass of adipose tissue on the dorsal side of the corpus callosum of our patient was neuroradiologically diagnosed as a PCL. Zetter et al. hypothesized early on that PCLs probably occur as a result of impaired gene expression [2]. More recently, it was reported that PCLs occasionally occur in association with impaired gene expression, such as with Pai syndrome [6,17] and anterior nasal dysplasia [16]. This suggests that PCL might be closely associated with gene expression disorder that extend not only within the skull but also beyond the skull.

The theory that PCL formationoccurs in the mesenchymal cell population of the meninx primitiva is also now widely accepted [3,9]. This cell population is formed as a tissue that supplies nutrients to cells during the early embryonic stage when the vegetative blood vessels have not yet formed, and are observed on the cranial side from the lamina terminalis to the area surrounding the choroid plexus until approximately the fifth to eighth weeks of the embryonic period [8].Wassermann's report [5] states that in the meninx primitiva, at approximately the fouth week of the embryonic period when angiogenesis initiated, the reticulated mesenchymal cells surrounding blood vessels remain undifferentiated and store fat to become adipocytes. It is speculated that abnormal gene expression in these reticulated mesenchymal cells may be the cause of PCL and excessive proliferation of adipocyte.

In addition to the symmetrical lipoma, our patient also exhibited very symmetrical calcification and brain malformations. A literature review of calcification similar to that in our patient stated that several cases of PCL are reported to

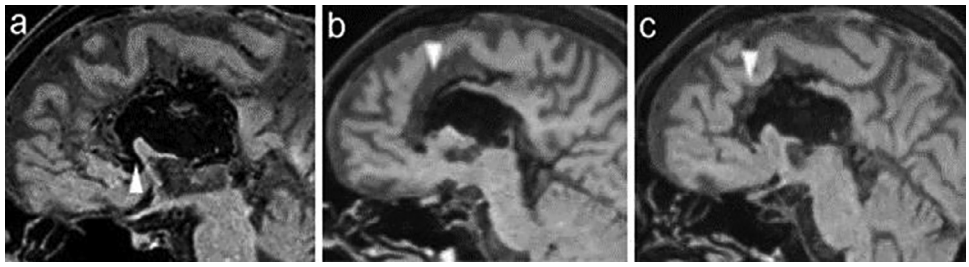

Fig. 5 – Sagittal fat-saturated T1-weighted magnetic resonance image of hypoplasia of the corpus callosum (A), which was only formed the rostrum (white arrow), and malformation of the right (B) and left (C) hemispheres. The first half of the cingulate gyrus on both sides was equally missing (white arrow).

have symmetric calcifications called bracket signs or curvilinear calcifications [7,10,15]. The presence of multiple previous cases with morphologically similar characteristics suggests a common background. This clearly differs from disordered neoplastic changes and may involve some sort of ordered mechanism. This can be explained by the theory [4] that organisms with 3 germ layers, including humans, develop with morphological symmetry from approximately the fourth week of the embryonic period. Further, the mesenchymal cell population is a group of cranial neural crest cells that have undergo epithelial-mesenchymal transition [13] from the neural crest in the early embryonic period. They have been shown to differentiate into the head and neck components, such as the facial bones, cartilage, nerve cells, and connective tissue [11]. It has also been reported, although in non-humans, that each of these cell groups migrates to a position determined by the induction of a specific factor, to form the body organs [1,11].This may be the mechanisms behind the symmetrical morphology of lipomas, calcifications and brain malformations as in our case.

The other features of our case were that the cingulate and paraterminal gyri contained excessive sheet-like adipose tissue formation. The dorsal side of the corpus callosum and part of the cingulate and paraterminal gyri anatomically correspond to indusium griseum and it is contained the arachnoid. In this regard, from Wassermann's report, we speculate that it also applies to the formation of excessive adipose tissue in the cingulate and paraterminal gyri. That is, in curvilinear type, which is another type of PCL is forming an excessive sheet-like adipose tissue morphology, an indusium griseum, which is an arachnoid of the same sheet-like morphology, was involved in its formation.

Regarding the cerebrovascular findings, the running of the left and right internal carotid arteries and their peripheral blood vessels are altered by the PCL and the surrounding excess adipose tissue. However in our case, the internal carotid artery normally branches into the pericallosal and callosomarginal arteries,passing towards the frontal and parietal lobe. Therefore, we concluded that there were at least no abnormalities in the development of the major blood vessels.

As mentioned above, the lesions in our case appear to have begun to form as early as fourth week after the embryonic period. In addition, it was presumed to be an expression abnormality caused by impaired gene that was present in a group of symmetrically migrating cranial neural crest cell populations. Therefore, it was speculated to be neurocristopathy [14].

Finally, regarding treatment. Many patients with PCL are asymptomatic. Tumors grow so slowly, therefore rarely indicated for neurosurgical intervention. Our case also did not display surgical indications. On the other hand, when the Monro foramen is occluded by the compression of the lipoma, the intracranial pressure may increase and the operation may be performed.

In summary, we reported a case with a symmetrical lesion in the midline of the skull. It was important that there were no symptoms and more than 80 years had passed. In the background, the relationship with the cranial neural crest cells seen in the early stage of development was speculated. Evaluation of the morphological symmetry of the lesion was considered to be a one of reference for prognosis.

Patient consent

Ethics approval and consent to participate

According to local ethical guidelines, all blood samples were obtained for storage and analysis only after written informed consent had been obtained from the patient. Written informed consent was obtained using a form approved by the local ethics committee (the Ethical Review Board of Daido Central Hospital [no.00032]). All procedures followed are accordance with the ethical standards of the responsible committee on human experimentation at Fukushima Medical University and with the 1975 Declaration of Helsinki, as revised in 2000.

Consent for publication

The patient agreed that the uses of his material may include(without limitation)publication of the materiali n the print and electronic editions of Radiology of Case Reports, on websites,sublicensed or reprinted editions(including foreign language editions),and in other works or products. Written informed consent was obtained from patient for publication of this case report any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

Availability of data and material

The datasets used and/or analyzed during the current study are available from the corresponding author on request.

Declaration of Competing Interest

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

REFERENCES

[1] Osborn AG. Anomalies of skill and meninges. Osborn's Brain: Imaging, Pathlogy, and Anatomy. Lippincott Williams & Wilkins; 2012. pp. 1202-1208.

[2] Zetter A, Netsky MG. Lipoma of the corpus callosum. J Neuropathol Exp Neurol 1950;19:305–19. doi:10.1097/00005072-196004000-00010.

[3] Truwit CL, Bankovich J. Pathogenesis of Intracranial Lipoma: an MR study in 42 Patients. Am J Roentgenol 1990;155(4):855–64. doi:10.2214/ajr.155.4.2119122.

[4] Matus DQ, Pang K, Marlow H, Dunn CW, Thomsen GH, Martindale MQ. Molecular evidence for deep evolutionary roots of bilaterality in animal development. Proc Natl Acad Sci U S A 2006;103(30):11195–200. doi:10.1073/pnas.0601257103.

[5] Wassermann F. Die Fettorgane des Menschen. Entwicklung, Bau und systematische Stellung des sog. Fettgewebes. Zellforsch 1926;3(235).

[6] Spai G, Levkoff AH, Jr Leithiser RE. Median cleft of the upper lip associated with lipomas of the central nervous system and cutaneous polyps. Am J Med Genet 1987;26(4):921–4. doi:10.1002/ajmg.1320260421.

[7] Taveras JM, Wood EH. Diagnostic Neuroradiology. Williams & Wilkins Co.; 1964. p. 1485–90.

[8] Osaka K, Handa H, Matsumoto S, Yasuda M. Development of the cerebrospinal fluid pathway in the normal and abnormal human embryos. Childs Brain 1980;6(1) pp. 26C-38. doi:10.1159/000119881.

[9] Krainer L. Die hirn-und ruckenmarkalipome. Virchew Arch Path Anat 1935;295:107–42. doi:10.1007/BF01888316.

[10] Erbay MF, Tecsllioglu M. A pericallosal lipoma case with abnormal vasculature mimicking arteriovenous malformation. Neurol India 2019;67(5):1331–3. doi:10.4103/0028-3886.271237.

[11] Minoux M, Holwerda S, Vitobello A, Kitazawa T, Kohler H, Stadler MB, et al. Gene bivalency at Polycomb domains regulates cranial neural crest positional identity. Science 2017;355(6332) pp.eaal2913. doi:10.1126/science.aal2913.

[12] Yilmaz N, Unal O, Kiymaz N, Yilmaz C, Etlik O O. Intracranial lipomas - a clinical study. Clin Neurol Neurosurg 2006;108(4):363–8. doi:10.1016/j.clineuro.2005.04.003.

[13] Francis-West PH, Robson L, Evans DJ. Craniofacial development: the tissue and molecular rtherinteractions that control development of the head. Adv Anat Embryol Cell Biol 2003;169(IIIVI):1–138. doi:10.1007/978-3-642-55570-1.

[14] Bolande RP. The neurocristopathies: A unifying concept of disease arising in neural crest maldevelopment. Hum Pathol 1974;5(4):409–29 (75)80115-8. doi:10.1016/s0046-8177.

[15] Gerber S, Plotkin R. Lipoma of the corpus callosum. Case report. J Neurosurg 1982;57(2):281–5. doi:10.3171/jns.1982.57.2.0281.

[16] Twigg SRF, Versnel SL, Numberg G, Lees MM, Bhat M, Hammond P, et al. Frontorhiny, a distinctive presentation of frontonasal dysplasia caused by recessive mutations in the ALX3 homeobox gene. Am J Hum Genet 2009;84(5):698–705. doi:10.1016/sjhg.2009.04.009.

[17] Rudnik-schoneborn S, Zerres K. A further patient with Pai syndrome with autosomal dominant inheritance? J Med Genet 1994;31(6):497–8 1994.PMID:8071979.