Journal of Neurosurgery Imaging and Techniques
JNSIT, 1(1): 1-6
www.scitcentral.com

Original Case Report: Open Access

Intracranial Extraskeletal Myxoid Chondrosarcoma of the Pineal Region in a Child and Literature Review

Shaun D. Rodgers, M.D., Nitin R. Wadhwani, M.D. and Tadanori Tomita*, M.D.

Division of Pediatric Neurological Surgery, and Department of Pathology, Ann & Robert H. Lurie Children's Hospital of Chicago, and Northwestern University Feinberg School of Medicine, Chicago, Illinois, USA

Received Nov 11, 2015; Accepted Jan 25, 2016; Published Apr 30, 2016

ABSTRACT

There have been less than a dozen reported cases of intracranial extraskeletalmyxoid chondrosarcoma in the literature. We present the youngest case to date with review of the literature. A 16-year-old boy initially presented with headaches and vomiting. At outlying hospital, the patient was found a large tumor in the pineal location with hydrocephalus. He was referred to our center after an intracranial biopsy following an endoscopic third ventriculostomywas not conclusive for pathology. He underwent resection at our institution and the pathology confirmed an extraskeletalmyxoid chondrosarcoma. The patient underwent adjuvant treatment, but ultimately required shunting for diffuse leptomeningeal disease. The shunt failed due to viscous CSF due to excessive presence ofmyxoid material. His disease progressed significantly and developed carcinomatous ascites and expired 16 months after diagnosis.

Keywords: Chondrosarcoma, Extraskeletal, Brain Tumor, Pineal Region, Children

INTRODUCTION

The first report of intracranial extraskeletalmyxoid chondrosarcoma was in 1972 by Enzinger [1]. There have been less than an dozen reports of this rare malignancy since then [1-10]. The tumors may be derived from the durain such locations as the falx or in the pineal region. It is thought that the tumor originates from the dura, leptomeninges, parenchyma, or the choroid plexus [9,11-15). We report the youngest patient in the literature with an intracranial myxoid chondrosarcoma which was located in the lateral ventricle.

Case Report

A 16-year-old male with no past medical history in otherwise normal health developed progressive headache over 2 months in the right frontal region. These were worse in the morning, but progressed to headache throughout the day. Additionally, he developed a few episode of emesis for 4-6 weeks. These symptoms prompted a CT and MRI of the brain at an outside institution. CT brain revealed a hypodense mass with patchy hyperdense lesion within it in the right thalamus to pineal region with obstructive hydrocephalus (**Figure 1**). MRI of the brain showed heterogeneous enhancing lesion extending to the quadrigeminal cistern and lateral ventricle from the right thalamus (**Figure 2**). He had no neurological deficits upon initial presentation.

The patient underwent endoscopic third ventriculostomy (ETV) and craniotomy at the outside hospital. The sample had a mucinous mucoid appearance. The pathology was inconclusive as the craniotomy was aborted due to brain swelling and only biopsy was obtained. His initial post-operative course was complicated by severe brain edema managed with induced coma. He made a substantial recovery, but was left with an upper quadrant hemianopia on the left side and short term memory dysfunction.

Corresponding author: Tadanori Tomita, M.D., Division of Pediatric Neurosurgery, Ann & Robert H. Lurie Children's Hospital of Chicago and 225 E. Chicago Avenue, Box 28 Chicago, Illinois 60611; Tel: 312.227.4220, FAX: 312.227.9679

Citation: Rodgers S, Wadhwani N & Tomita T (2016) Intracranial Extraskeletal Myxoid Chondrosarcoma of the Pineal Region in a Child and Literature Review. J Neurosurg Imaging Techniques, 1(1): 1-6.

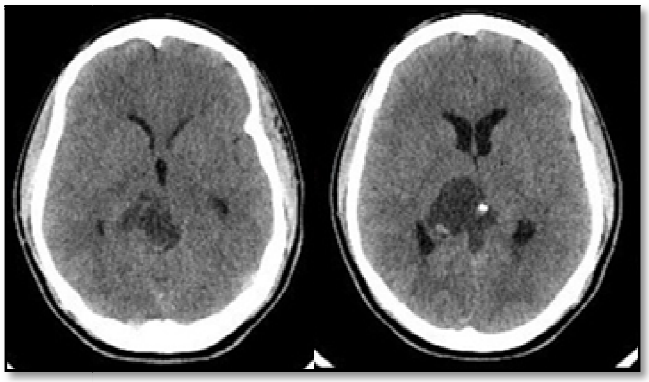

Figure 1. Non-ciontrast axial head CT showing a hypodense mass lesion in the right thalamus extending to the pineal region.

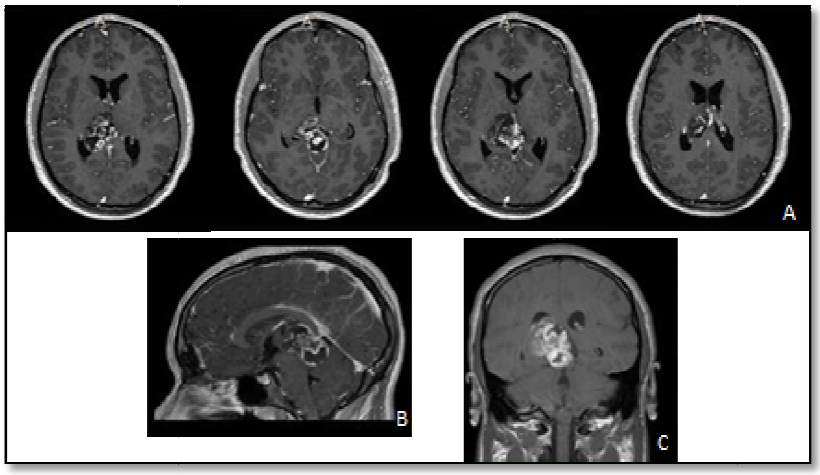

Figure 2. Postcontrast MR, axial (A), sagittal (B) and coronal (C) images showing a heterogeneously enhancing mass lesion in the right thalamus, lateral ventricle and pineal region

Over the next month, the tumor continued to rapidly increase in size and cross the midline. There was cerebral herniation of the occipital lobe out the craniotomy site as well. MR showed contrast enhancing large tumor in the pineal location extending to the lateral ventricle involving the thalamus of the right side (**Figure 3**). His examination showed a left-side homonymous hemianopia and Parinaud signs. Four months after the initial craniotomy, he was transferred to our institution for a second opinion and elected for a repeat craniotomy for further tumor resection. At the time of surgery an external ventricular drain (EVD) was placed for brain relaxation. He underwent an occipital transtentorial approach for sub-total resection of the tumor (**Figure 4**). Tumor resection was stopped after extensive debulking due to the involvement of the vein of Galen and internal cerebral veins. Post-operatively he underwent a ventriculo-peritoneal (VP) shunt placement 1 week after resection due to inability to wean off the EVD .The CSF profiles at that time showed protein 50 mg/dL, glucose61mg/dL, 24rbc /cu.mm and 1 wbc/cu.mm without tumor cells. His Parinaud syndrome eventually resolved.

The pathology revealed individual neoplastic cells floating in an abundant amount of myxoid material. Some of these individual cells formed small groups or cord-like structures, and a majority of the cells had a bubbly to vacuolated cytoplasm (**Figure 5A, 5B**). The tumor stained strongly for Vimentin (**Figure 5C**). It had occasional cells positive for EMA, SMA, S-100, and Calponin. D2-40, GFAP, C-Kit, Keratin AE1/AE3, and P63 were negative. There was loss of normal nuclear expression of INI-1 (**Figure 5D**). The differential diagnosis included an unusual Atypical Teratoid Rhaboid Tumor, Myoepithelial Carcinoma, and an Extraskeletal Myxoid Chondrosarcoma. Occasional interspersed fragments of dense, disordered hyaline cartilage with atypical appearing chondrocyte-like cells and calcification were seen. In conjunction with the above immunohistochemical stains, the morphology favored an Extraskeletal Myxoid Chondrosarcoma.

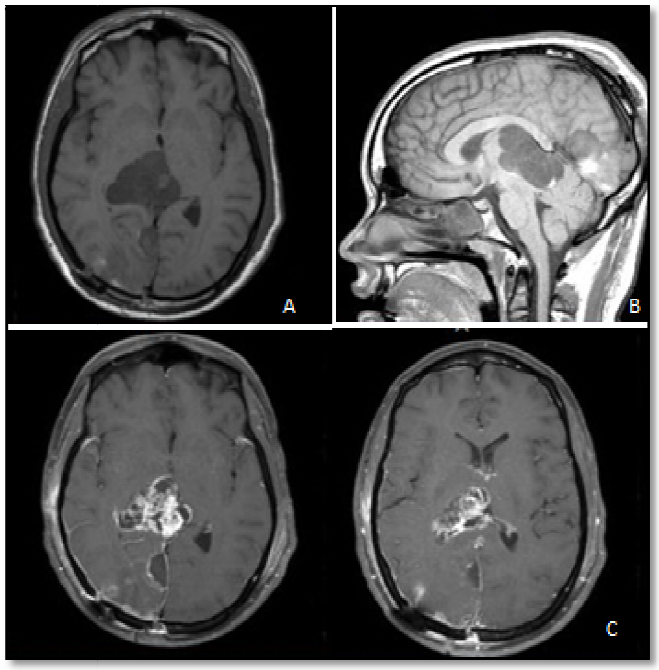

Figure 3. MRI of pineal extraskeletalmyxoid chondrosarcoma after recurrence **.** Pre contrast T-1 weighted axial (A) and sagittal (B) images, and post contrast axial images (C)

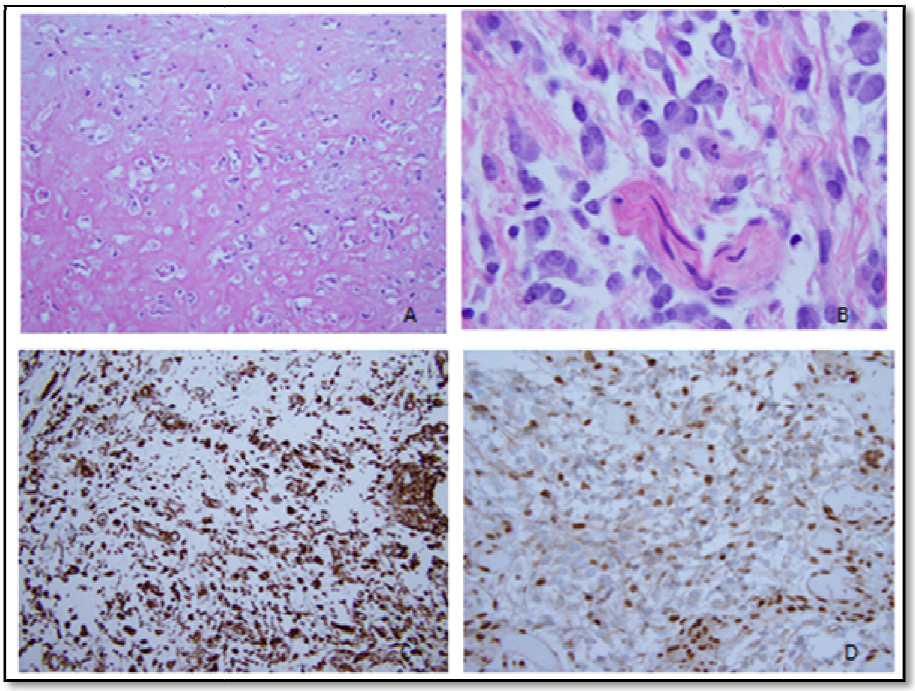

Figure 4. Photomicrography of extraskeletalmyxoid chondrosarcoma. The pathology revealed individual neoplastic cells floating in an abundant amount of myxoid material and disordered hyaline cartilage.. Some of these individual cells formed small groups or cord-like structures, and a majority of the cells had a bubbly tovacuolated cytoplasm (Figure 3 A x100; B x400). The tumor stained strongly for Vimentin (**Figure 3 C**). It had occasional cells positive for EMA, SMA, S-100, and Calponin. D2-40, GFAP, C-Kit, Keratin AE1/AE3, and P63 were negative. There was nuclear loss of INI-1 (**Figure 3 D**).

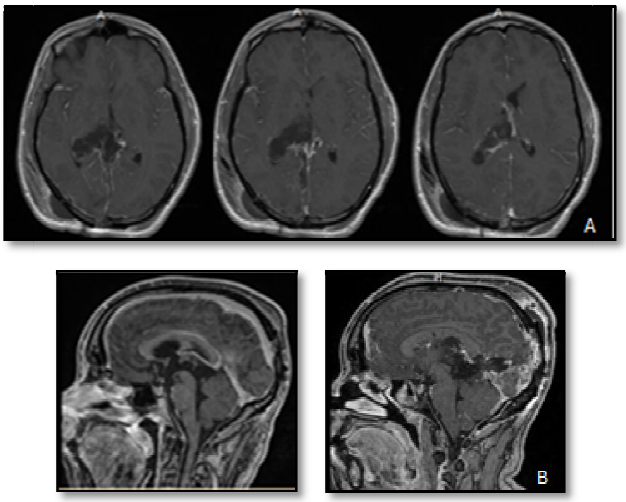

Figure 5. Postcontrast axial (A) and sagittal (B) MR images after subtotal resection of extraskeletalmyxoid chondrosarcoma through occipital transtentorial approach

He underwent adjuvant treatment with proton radiation therapy and pazopanib. He received craniospinal irradiation with boost to the original tumor area due to positive cerebrospinal fluid (CSF) cytology. He had boost of 30.67 cobalt grey equivalent (CGE) to the primary site and 36.30 CGE to the craniospinal axis at 2 months postoperatively. However, over the next 6 months he developed multiple cranial nerve abnormality. By 12 months postoperatively, he had decreased vision bilaterally, bilateral hearing loss, facial paralysis, and uvula deviation. His MRI brain revealed stable lateral ventricle and thalamic disease with subtle leptomeningeal enhancement .

One month later, he underwent VP shunt exploration after his lateral ventricles became large and he became poorly responsive (**Figure 6**). The endoscopic observation showed open ETV but whitish clumps of tumor floating between the ventricle and prepontinesubarachnoid space, and the ventricular CSF was extremely viscous (**Video 1**). The CSF was extremely viscous. The CSF profiles indicate protein 126 mg/dL, glucose 50 mg/dL, 71 red blood cell /cu.mm and 7 white blood cell/cu.mm with tumor cells. His abdomen was not absorbing the fluid causing extensive ascites. The CSF profile 10 days later showed protein 275 mg.dL, glucose 68 mg/dL, 275 rbc/cu.mm and 52 wbc/cu.mm. Both CSF and ascites cytology revealed extensive malignancy. He ultimately received an external pigtail catheter placement into the peritoneum for the ascites and was discharged. He expired 16 months after the diagnosis.

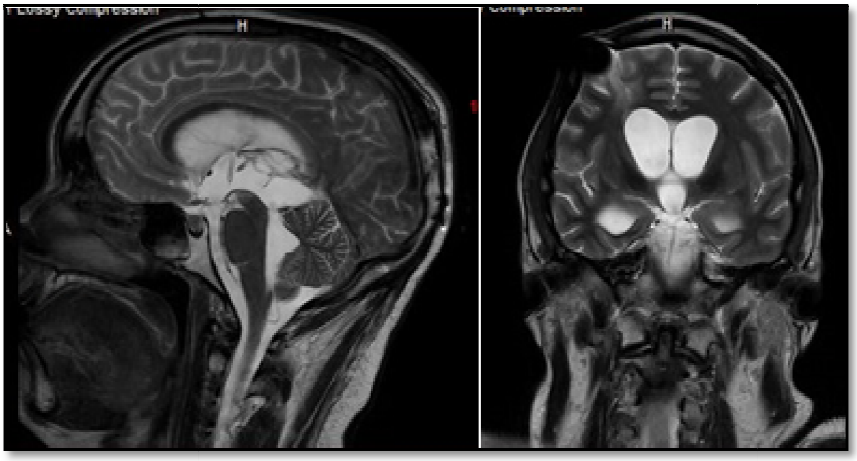

Figure 6. Fast T2 weighted MR shows enlarged ventricles.

Video 1. Neuroendoscopic view of the right lateral ventricle. The endoscope passing through the lateral ventricle to Pthe third ventricle. Patent third ventriculostomy is noted at the floor of the third ventricle. Note whitish clumps of tumor floating between the ventricle and prepontine subarachnoid space. The ventricular CSF flowing out the catheter was viscous like syrup.

DISCUSSION

Intracranial extraskeletalmyxoid chondrosarcoma is extremely rare. Extraskeletalmyxoid chondrosarcoma has been described in many organ systems [1,16,17]. Our case appears to have originated from the choroid plexus, thalamus, or the pineal region. A similar case of thalamic location as ours was reported by Park et al. [18], which was considered to have originated from the choroid plexus. However, the extensive tumor invasion makes the origin hard to determine. Our case did not have significant calcification as others have described [16]. The S-100 was focally present as previously described [16,17]. The INI-1 loss is still unclear for the diagnostic utility [19]. However, INI1 negative extraskeletalmyxoid chondrosarcoma was reported [20].

Importance of total resection was emphasized for tumor control. However, complete resection is often not possible for those occurring in the cerebellopontine angles, pineal and sellar location. In our case a gross total resection was not attainable with the extensive invasiveness. Park et al. reviewed the 8 cases reported in the literature [18]. Six of these eight were able to have a complete resection. It appears the aggressive nature of the tumor warrants an aggressive, but safe surgical plan. Radiation therapy seems appropriate for such a highly malignant tumor [2,3,5,7,10]. However, it is unclear about a survival advantage with adjuvant chemotherapy.

At the terminal stage, the patient showed widespread metastasis in the ventricle and subarachnoid space together with peritoneal cavity through the shunt. The viscous nature of the ventricular CSF is due to not only the elevated CSF protein and numerous malignant cells in the CSF but also to the presence of mucin secreted by myxoidchondrosarcoma.

CONCLUSION

Our case is the youngest reported of an intracranial extraskeletalmyxoid chondrosarcoma. There is a paucity of available data in the literature for these rare tumors. We advocate aggressive maximal safe resection with upfront adjuvant therapy. One should be aware extremely viscous CSF due to presence of myxoid material once the tumor disseminates in the cerebral ventricle.

REFERENCES

1. Enzinger FM, Shiraki M (1972) Extraskeletalmyxoid chondrosarcoma. An analysis of 34 cases. Hum Pathol 3: 421-435.
2. Chaskis C, Michotte A, Goossens A, Stadnik T, Koerts G, et al. (2002) Primary intracerebral myxoid chondrosarcoma. Case illustration. J Neurosurg 97: 228.
3. Gonzalez-Lois C, Cuevas C, Abdullah O, Ricoy JR (2002) Intracranial extraskeletalmyxoid chondrosarcoma: case report and review of the literature. ActaNeuorochir (Wien) 144: 735-740.
4. Hero RC, Martinez AJ, Ahn HS (1980) Intracranial mesenchymal chondrosarcoma. Surg Neurol 14: 311-317.
5. Im SH, Kim DG, Park IA, Chi JG (2003) Primary intracranial myxoid chondrosarcoma: report of a case and review of the literature. J Korean Med Sci 18: 301-307.

6. Sala F, Talacchi A, Beltramello A, Izzulino P, Bricolo A (1998) Intracranial myxoid chondrosarcoma with early intradural growth. J Neurosurg Sci 42: 159-163.
7. Salcman M, Scholtz H, Kristt D, Numaguchi Y (1992) Extraskeletalmyxoid chondrosarcoma of the falx. Neurosurgery 31: 344-348.
8. Sato K, Kubota T, Yoshida K, Murata H (1993) Intracranial extraskeletalmyxoid chondrosarcoma with special reference to lamellar inculusions in the rough endoplasmic reticulum. Acta Neuroplathol 86: 525-528.
9. Scott RM, Dickersin R, Wolpert SM, Twitchell T (1976) Myxochondrosarcoma of the fourth ventricle. Case report. J Neurosurg 44: 386-389.
10. Sorimachi T, Sasaki O, Nakazato S, Koike T, Shibuya H (2008) Myxoid Chondrosarcoma in the pineal region. J Neurosurg 109: 904-907.
11. Bourgouin PM, Tampieri D, Robitaille, Y, Robert F, Bergeron D, et al. (1992) Low-grade myxoid chondrosarcoma of the base of the skull: CT, MR, and histopathology J Comput Assit Tomogr 16: 268-273.
12. Cummings TJ, Bridge JA, Fukushima T (2004) Extraskeletalmyxoid chondrosarcoma of the jugular foramen. Clin Neuropathol 23: 232-237.
13. Grossman RI, Davis KR (1981) Cranial computed tomographic appearance eof chondrosarcoma of the base of the skull. Radiology 141: 403-408.
14. Hassounah M, Al-Melfty O, Akhtar M, Jinkins JR, Fox JL (1985) Primary cranial and intracranial chondrosarcoma. A survey. ActaNeurochir(Wien) 78: 123-132.
15. Smith TW, Davidson RI (1981) Primary meningeal myxochondrosarcoma presenting as a cerebellar mass: case report. Neurosurgery 8: 577-581.
16. O'Brien J, Thornton J, Cawley D, Farrell M, Keohane K, et al. (2008) Extraskeletalmyxoid chondrosarcoma of the cerebellopontine angle presenting during pregnancy. Br J Neurosurg 22: 429-432.
17. Oliveira AM, Sebo TJ, McGrory JE, Gaffey TA, Rock MG, et al. (2000) Extraskeletalmyxoid chondrosarcoma: a clincopathologic, immunohistochemical, and ploidy analysis of 23 cases. Mod Pathol 13: 900-908.
18. Park JH, Kim MJ, Kim JK, Kim JH (2012) Intracranial extraskeletalmyxoid chondrosarcoma: case report and literature review. J Korean Neurosurg Soc 52: 246-249.
19. Korten AG, ter Berg HJ, Spincemaille GH, van der Laan RT, Van de Wel AM (1998) Intracranial chondrosarcoma: review of the literature and report of 15 cases. J NeurolNeurosurg Psychiatry 65: 88-92.
20. Kohashi K, Oda Y, Yamamoto H, Tamiya S, Oshiro Y, et al. (2008) SMARCB1/INI1 protein expression in round cell soft tissue sarcomas associated with chromosomal translocations involving EWS: A special reference to SMARCB1/INI1 negative variant extraskeletalmyxoid chondrosarcoma. Amr J Surgical Pathology 32: 1168-1174.