

Case Report

Cicatricial Fibromatosis Mimics Metastatic Medulloblastoma

Kouichirou Okamoto, Jusuke Ito, and Kunio Sakai

Summary: Cicatricial fibromatoses usually occur in the anterior abdominal wall or in the extremities, but rarely in the scalp or the soft tissues of the neck. We report a case of desmoid fibromatosis that developed in a 15-year-old boy 8 months after surgery for cerebellar medulloblastoma.

The fibromatoses are distinctive lesions best defined as a group of nonmetastasizing fibrous tumors that tend to invade locally and recur after surgical excision (1). Desmoid fibromatosis is a synonym for deep fibromatosis, and is one of the adult fibromatoses, which occur predominantly but not exclusively in patients older than 20 years (1).

Desmoid fibromatosis that arises in a surgical scar, cicatricial fibromatosis, is a well-known clinical condition (1, 2); however, its occurrence after neurosurgery in the scalp and soft tissues of the neck is extremely rare. We report a case of desmoid fibromatosis that developed 8 months after surgery for cerebellar medulloblastoma, in which differentiation from extraneural metastasis posed a clinical problem.

Case Report

A small tumor under the right semispinalis capitis muscle was found on a follow-up MR examination in a 15-year-old boy. He had undergone surgery 8 months earlier for cerebellar medulloblastoma and had received craniospinal irradiation postoperatively. The tumor, which was not seen on a CT scan obtained 4 months earlier, was under the scar of the midline suboccipital skin incision and was isointense with overlying muscle on T1-weighted MR images (Fig 1A) and homogeneously isointense relative to fat on T2-weighted images (Fig 1B). It enhanced markedly and homogeneously after intravenous injection of gadopentetate dimeglumine (Fig 1C). The tumor was round on axial images but fusiform on sagittal images. There was no imaging findings of recurrence or dissemination of medulloblastoma in the CNS.

Although extraneural metastasis of cerebellar medulloblastoma without recurrence and dissemination in the CNS is a rare clinical condition, metastasis was considered as a possible diagnosis. The histopathologic diagnosis of desmoid fibromatosis was made on a surgically excised specimen.

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From the Departments of Radiology and Dentistry, Niigata University Schools of Medicine, Niigata, Japan.

Address reprint requests to K. Okamoto, MD, Department of Radiology, Niigata University School of Medicine, 1-757 Asahimachi-dori, Niigata, 951-8510 Japan.

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Discussion

Desmoid fibromatosis is one of the adult fibromatoses and occurs predominantly in patients older than 20 years, although it can occur in children (1). Desmoid fibromatosis arising in a surgical scar is well known as cicatricial fibromatosis (2). Cicatricial fibromatoses usually occur in the anterior abdominal wall or the extremities (1); they are extremely rare in the scalp and soft tissue of the neck after surgery (2).

Extracranial metastases from primary intracranial tumors are rare occurrences. However, extraneural metastases of brain tumors occur frequently in patients with cerebellar medulloblastoma (3-6). Bone and bone marrow are the most commonly involved sites, followed by the lymph nodes (3, 6), but metastases to other sites, including the peritoneum, liver, lung, pancreas, and breasts, have also been reported (3-6). Extraneural metastases can occur without local recurrence or dissemination within the CNS and without clinical manifestations of any other metastatic foci (5, 6).

MR findings of desmoids are nonspecific and are variable in configuration and in signal intensity on MR images (7, 8). However, desmoids are usually hypointense or isointense relative to muscle on T1-weighted images, and show moderate or marked contrast enhancement in most cases. On T2-weighted images, smaller desmoids are homogeneously hyperintense, as seen in our case (7, 8). Therefore, MR characteristics of desmoid fibromatosis may be similar to those of metastatic medulloblastoma.

The average survival time for patients with systemic metastases from cerebellar medulloblastoma is shorter than that for patients without systemic metastases (3), and the prognosis is almost uniformly fatal (4). Therefore, differentiating cicatricial fibromatosis from extraneural metastasis is clinically important, but it was difficult in our case. Retrospectively, cicatricial fibromatosis is the more likely finding on MR images because of the characteristic location of the tumor and its fusiform configuration, with the long axis oriented in the direction of the covering muscle bundle.

Conclusion

Cicatricial desmoid fibromatosis in the scalp and soft tissues of the neck is an extremely rare clinical condition; however, it should be included in the

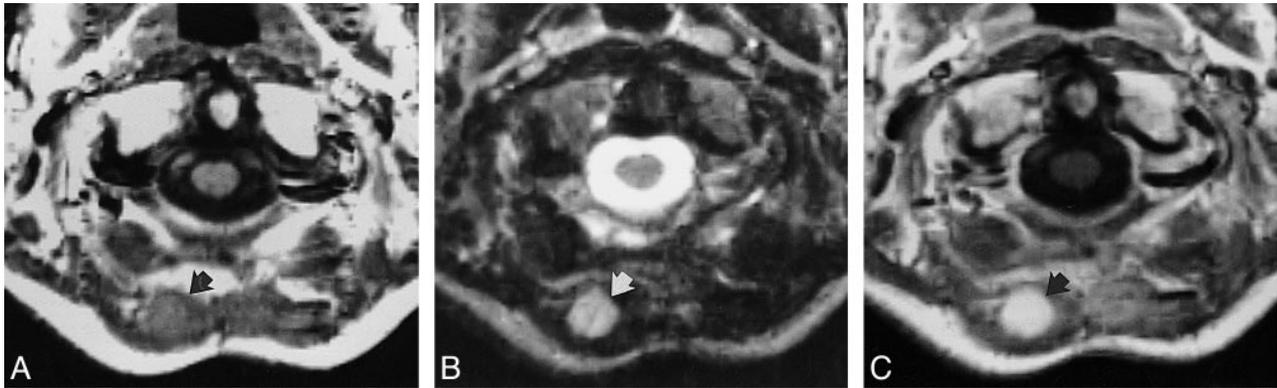


FIG 1. Axial spin-echo MR images in a 15-year-old boy acquired 8 months after surgery for cerebellar medulloblastoma.

A, T1-weighted image (600/15/2 [TR/TE/excitations]) shows an isointense small round mass (*arrow*) under the right semispinalis capitis muscle.

B, On T2-weighted image (3000/90/1), the mass (*arrow*) is well demarcated and isointense relative to subcutaneous fat.

C, On postcontrast T1-weighted image (600/15/2), the mass (*arrow*) enhances markedly and homogeneously.

differential diagnosis of a tumor arising in the scar after neurosurgery.

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