A Bednar tumor is a rare neoplasm of intermediate malignant potential that accounts for 1-5% of all cases of dermatofibrosarcoma protuberans (DFSP). This tumor is considered a pigmented variant of DFSP, because the clinical and histological findings resemble DFSP. The diagnosis is commonly made in early to middle adult life except in cases with melanin containing cells. In the case presented here, the patient was a 3-year-old male who presented with a painless slow-growing 2.0 × 1.5 × 1.0 cm mass on the dorsal aspect of his right hand. Histological examination of the biopsy specimen revealed typical features of a Bednar tumor, which was composed of CD34 positive monomorphous spindle shaped cells arranged in a storiform fashion with moderate mitotic activity (up to 5 per 10 HPF) and scattered pigmented cells with dendritic processes. We report a rare case of Bednar tumor affecting a pediatric patient and review the medical literatures.

Key Words: Dermatofibrosarcoma; Pigmentation; Pediatrics

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A Bednar tumor is a rare neoplasm of intermediate malignant potential. It accounts for 1-5% of all cases of dermatofibrosarcoma protuberans (DFSP). In 1957, Bednar first described a group of nine cutaneous tumors that were characterized by indolent growth and a prominent storiform pattern, and in four cases by the presence of melanin pigment. He regarded these tumors as a variant of neurofibromas (storiform neurofibroma). However, this tumor is currently considered a pigmented variant of DFSP due to histological and cytogenetic similarities between the two lesions. Most of the cases with this type of tumor described to date are adults; there are very few reports of a Bednar tumor affecting pediatric patients.

We report a rare case of a Bednar tumor in a 3-year-old patient and review the literature.

CASE REPORT

A 3-year-old previously healthy male presented with a slow-growing painless soft tissue mass on the thumb of the right hand. An ultrasound examination showed a heterogeneous hypoechoic mass in the subcutis at the fifth metacarpal area of the right hand (Fig. 1). Macroscopic examination revealed several irregular fragments of firm, brownish-black or grayish-white tissue, measuring 2.0 cm in the greatest dimension.

Light microscopic examination of hematoxylin-eosin stained sections, of a biopsy specimen, showed a tumor composed of monomorphous spindle shaped cells arranged in a prominent storiform or cartwheel fashion with infiltration (Fig. 2A-C). These cells showed medium-sized, slightly atypical but monotonous hyperchromatic nuclei and amphophilic to eosinophilic cytoplasm with poorly defined cell borders. Moderate mitotic activity (up to 5/10 HPF) was noted. There were scattered heavily pigmented cells with round to oval vesicular nuclei and dendritic cytoplasm in the spindle cells. These pigmented cells stained with Fontana-Masson stain (Fig. 2D).

Immunohistochemically, the spindle tumor cells were diffusely positive for vimentin (Zymed, San Francisco, CA, USA, dilution 1:50) and CD34 antigen (Novocastra, Newcastle, UK, dilution 1:50) (Fig. 3A). They were negative for all other markers, in-
cluding keratin AE1:AE3 (Dakocytomation, Glostrup, Denmark, dilution 1:100), S-100 protein (Dakocytomation, dilution 1:100), HMB-45 (Dakocytomation, dilution 1:100), CD56 (Zymed, dilution 1:50), neuron specific enolase (Zymed, dilution 1:50), smooth muscle actin (Dakocytomation, dilution 1:100), desmin (Zymed, dilution 1:50) and factor VIII-related antigen (Dakocytomation, dilution 1:30). The pigmented cells were positive for S-100 protein (Fig. 3B), HMB-45 (Fig. 3C), and CD56, but negative for all other markers.

Thus, the diagnosis of Bednar tumor (pigmented DFSP) was

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**Fig. 1.** High frequency ultrasonogram examination shows a heterogeneously hypoechoic mass (arrows) in subcutaneous fat layer of the thumb (A). Color-Doppler application reveals peripheral vascularity in the mass. Some hyperechogenic areas with no enhancement (open arrow) are also noted (B).

**Fig. 2.** The tumor is composed of monomorphous spindle shaped cells arranged in prominent storiform or cartwheel fashion and scattered pigmented cells with dendritic process (A). The tumor cells infiltrate between adnexal structures and normal adipocytes (B & C). The pigmented dendritic cells are positive for Fontana Masson stain (D).
made. The 8 month follow-up period was uneventful.

**DISCUSSION**

Bednar tumors differ from the typical cases of DFSP by the presence of heavily melanin pigmented dendritic cells, the histiogenesis of which continues to be debated. Several investigators have suggested that this tumor is derived from neuroectodermal cells according to the ultrastructural and immunohistochemical findings as well as the presence of melanosome containing cells. In the present case, the pigmented dendritic cells were positive for S-100 protein and CD56. However, the CD34 positive non-pigmented spindle cells were completely negative for S-100 protein, CD56 and neuron specific enolase. These results suggest that the CD34 positive spindle cells and pigmented dendritic cells have a different origin. However, to date it is uncertain whether Bednar tumors are simply colonized by the melanin bearing cells or the tumor is derived from putative neuromesenchyme.

The differential diagnoses include other benign or malignant cutaneous pigmented neoplasms such as pigmented (melanotic) neurofibroma, psammomatous melanotic schwannoma, and desmoplastic (neurotrophic) melanoma. A pigmented neurofibroma can be confused with a Bednar tumor because the melanin-laden cells of both processes are similar. However, the Bednar tumor exhibits a more extensive storiform growth, has greater immunoreactivity for CD34 and lacks diffuse proliferation of S-100 protein positive Schwann cells. Psammomatous melanotic schwannoma is rather circumscribed, heavily pigmented with psammomatous bodies and diffusely positive for S-100 protein, whereas the Bednar tumor is poorly circumscribed and composed of CD34 positive spindle shaped cells with scattered pigmented cells. Desmoplastic (neurotrophic) melanoma shows a neurotropism, focal melanocytic junctional activity, and diffuse and strong S-100 protein immunoreactivity. In addition to a careful histology examination, immunohistochemical study for CD34 is the most useful marker for differentiating a Bednar tumor from other cutaneous pigmented tumors.

The typical DFSP has a rate of recurrence ranging from 20-50% among reports with a long-term follow up. There are reports of rare cases of Bednar tumors with distant metastasis. However, their biological behavior is usually less aggressive than the typical DFSP. Mochzuchi cited a recurrence rate of 17% among reported cases of Bednar tumors. In addition, they reported an average interval of 9 years for recurrence (range, 9 months to 23 years). Although it is difficult to assess the biologic behavior of this tumor in pediatric patients because of the rarity of cases, 3 cases with recurrence have been reported among the 6 cases of pediatric Bednar tumor. Table 1 summarizes the clinical data of the reported cases to date.

The recommended treatment for DFSP or Bednar tumors in the adult or pediatric patient is wide excision with more than 2-3 cm margins of visibly uninvolved tissue and inclusion of the superficial fascia. However, this wide excision is difficult in

**Table 1. Summary of the clinical data of reported pediatric Bednar tumors**

<table>
<thead>
<tr>
<th>Age/Gender</th>
<th>Ethnicity</th>
<th>Site</th>
<th>Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>1&quot;</td>
<td>6 months/F</td>
<td>Asian</td>
<td>Lower back</td>
</tr>
<tr>
<td>2&quot;</td>
<td>Congenital/F</td>
<td>Black</td>
<td>Lumbar area</td>
</tr>
<tr>
<td>3&quot;</td>
<td>2.5 years/F</td>
<td>Black</td>
<td>Vulva</td>
</tr>
<tr>
<td>4&quot;</td>
<td>12 years/M</td>
<td>White</td>
<td>Foot</td>
</tr>
<tr>
<td>5&quot;</td>
<td>7 years/F</td>
<td>Black</td>
<td>Shoulder</td>
</tr>
<tr>
<td>6&quot;</td>
<td>17 years/F</td>
<td>Asian</td>
<td>Back</td>
</tr>
<tr>
<td>7&quot;</td>
<td>3 years/M</td>
<td>Asian</td>
<td>Hand</td>
</tr>
</tbody>
</table>

*Present case, F; female, M; male, GCF; giant cell fibroblastoma.
cases of infants or early childhood patients, especially when present on the hand or foot. Therefore, closer clinical follow up should be recommended for early detection of local recurrence after a surgical excision in pediatric patients.

REFERENCES