

# Congenital giant common blue nevus of the scalp (A case report)

## Saçlı deride doğumsal dev yalın mavi nevüs (Olgu sunumu)

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### Summary

**Purpose:** Common blue nevi are melanocytic tumors rarely exceeding 1cm in diameter. In this paper, a case with a congenital giant common blue nevus of the scalp is presented and the literature about the giant blue nevi is reviewed.

**Case report:** A 31-year-old woman was admitted to hospital with a pigmented lesion on the occipital scalp that had been present since birth. Physical examination showed a 7.5x7cm blue-black lesion. After a biopsy was performed, complete surgical excision was attempted. After the histopathologic examination, the lesion was diagnosed as common blue nevus.

**Conclusion:** There are only four reported cases of congenital giant common blue nevi in the literature. Because of its rarity and the lack of long-term follow-up in reported cases, it is difficult to assess its behavior. Although the risk of recurrence or malignant transformation seems to be very low, long-term follow-up is recommended.

**Key words:** Common blue nevus, giant, melanocytic tumors

### Özet

**Amaç:** Yalın mavi nevüs'ler çapları 1 cm'yi çok az aşan melanositik tümörlerdir. Bu makalede, saçlı deride yerleşim gösteren doğumsal dev yalın mavi nevüs saptanan bir olgu sunulmuş ve dev mavi nevüs'lerle ilgili kaynak bilgileri gözden geçirilmiştir.

**Olgu sunumu:** 31 yaşındaki kadın hasta oksipital bölge saçlı derisinde doğumdan beri var olan pigmente lezyon nedeniyle başvurdu. Fizik bakıda, oksipital bölge saçlı derisinde 7.5x7 cm boyutlarında mavi-siyah renkli lezyon saptandı. Biyopsi ardından lezyon total olarak çıkarıldı. Histopatolojik inceleme sonucunda yalın mavi nevüs tanısı kondu.

**Sonuç:** Kaynaklarda doğumsal dev yalın mavi nevüs saptanan sadece dört olgu bildirilmiştir. Az oluşları ve sunulan olgularda uzun süreli izlem bilgilerinin bulunmaması nedeniyle bu lezyonların davranışını belirlemek zordur. Rekürrens ve malign transformasyon riski çok düşük olmakla birlikte, bu olgularda uzun süreli izlem önerilmektedir.

**Anahtar sözcükler:** Yalın mavi nevüs, dev, melanositik tümör

**B**lue nevi are pigmented lesions composed of spindle-shaped dendritic and/or oval dermal melanocytes (1). They generally occur on the buttocks, dorsum of the hands and feet, scalp, and face (2). Four types of blue nevi, designated as "common", "cellular", "combined" and "epithelioid", have been recognized. Blue nevi usually occurs as a small nodule less than 3 cm in diameter (3, 4). Giant blue nevi are exceptional. Herein we describe a patient with a giant common blue nevus of the scalp.

### Case report

A 31-year-old woman was seen with a pigmented lesion on the occipital scalp that had been present since birth. The size of the lesion remained unchanged until childhood when it began to enlarge slowly. Physical examination showed a 7.5x7 cm blue-black lesion. Incisional biopsy of the lesion revealed bundles of spindle-shaped melanocytes with melanin granules and melanophages near the bundles of melanocytes in the

dermis. These histopathologic findings were consistent with the diagnosis of blue nevus and thereafter complete surgical excision was performed. Surgical specimen showed a blue-black, dome-shaped tumor (Fig 1).

Microscopically, the lesion was composed of numerous spindle-shaped or dendritic, slightly wavy melanocytes filled with melanin granules involving the entire thickness of the dermis (Fig 2). After melanin bleaching, the lesion showed melanocytes that are devoid of mitoses and atypia. No necrosis was observed. There was also melanophages near the bundles of melanocytes and slight increase in fibrous tissue (Fig 3). The epidermis was normal.

### Discussion

The blue nevus is a pigmented lesion consisting of dermal melanocytes that can appear in diverse forms: dendritic, spindle-shaped, oval-shaped, or polyhedral (1). There are four histologic variants, designated as "common", "cellular", "combined" and "epithelioid". Among them, common blue nevi are flat or discretely raised lesion of dark blue color (1). Common blue nevi are composed of greatly elongated, slender melanocytes with long, occasionally branching dendritic processes that contain abundant melanin. These elongated melanocytes are often grouped in irregular bundles in the dermis, sometimes extending into the subcutis.

Numerous melanin-laden macrophages are often observed among the bundles of melanocytes. There is also extensive fibrosis. The epidermis is normal and junctional activity is consistently absent.

Cellular blue nevi in addition to the deeply pigmented dendritic melanocytes like those of common blue nevi, are composed of cellular islands of large oval to round melanocytes with abundant pale cytoplasm, containing little or no melanin. Melanophages may be present between these islands.

Epithelioid blue nevus is a rare variant of blue nevus that have been recently described mostly in patients with the Carney complex (5). Epithelioid blue nevus is composed of melanin-laden polygonal epithelioid melanocytes. In this variant, the neoplastic cells show no maturation at the base of the lesion and, in contrast with the usual stromal changes in blue nevi, epithelioid variant exhibits no dermal fibrosis.

In our case, histopathologic examination demonstrated bundles of spindle-shaped or dendritic melanocytes filled with melanin granules accompanied by numerous melanophages, features consistent with the diagnosis of "common blue nevus".

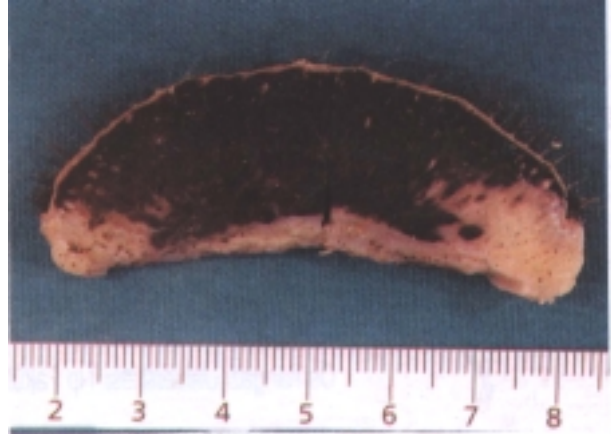


Figure 1. Surgical specimen showing a blue-black, dome-shaped lesion.

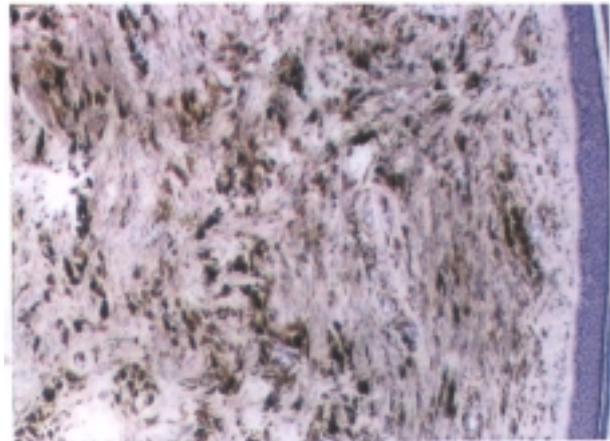


Figure 2. Spindle-shaped melanocytes filled with melanin granules in the entire thickness of the dermis (H+E stain; original magnification X 40).

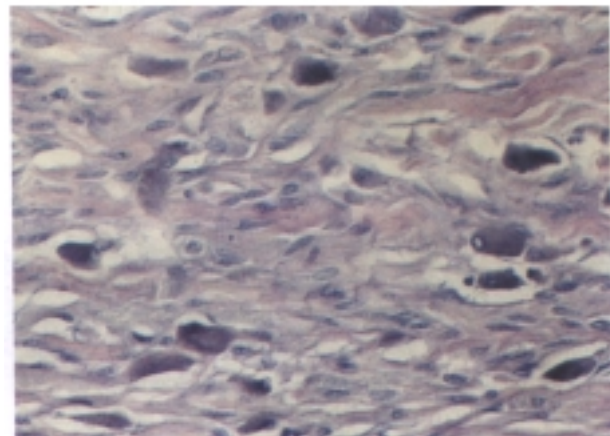


Figure 3. Spindle-shaped, slightly wavy melanocytes and melanophages in the dermis (H+E stain after melanin bleaching; original magnification X 200).

Blue nevi are small, usually acquired melanocytic tumors most often located over the extremities or scalp. The common blue nevi usually occurs as a small nodule rarely exceeding 1 cm in diameter (4). The cellular blue nevi are papulous, dome-shaped or flat lesions that are usually larger than the common blue nevus and generally measures 1 to 3 cm in diameter (4). Giant blue nevi are exceptional. There have been a few cases of giant blue nevi reported in the literature and most of them are cellular blue nevi (2,3,6-13). To our best knowledge, there have only been four cases of giant common blue nevi in the literature and all these cases were congenital (Table I). Upshaw et al. (14) described a 9-year-old child with a 17x6 cm lesion on the lateral aspect of thorax that had been seen at four weeks of age. The excision specimen revealed a blue nevus of Jadassohn-Tieche.

Pittman and Fisher (15) described an 18-year-old man with a 8x6cm common blue nevus on his left calf since birth. Radentz and Vogel (16) reported a 8-month-old girl with a congenital 7x5 cm lesion, consistent with a common blue nevus, over the right parietal aspect of her scalp. The fourth case, reported by Kawasaki et al. (17), was a 43-year-old man with a 6x5cm common blue nevus on the parietal scalp since birth.

Table I. Previously reported cases of congenital giant common blue nevus

Previous reports	Site	Age	Size
Upshaw et al. <sup>14</sup>	Thorax	9	17x6 cm
Pitmann and Fisher <sup>15</sup>	Left calf	18	8x6 cm
Radentz and Vogel <sup>16</sup>	Scalp	8/12	7x5 cm
Kawasaki et al. <sup>17</sup>	Scalp	43	6x5 cm

## References

- Gonzales-Campora R, Galera-Davidson H, Vasquez-Ramirez FJ, Diaz-Cano S. Blue nevus: Classical types and new related entities. A differential diagnostic review. *Path Res Pract* 1994; 190: 627-635.
- Hoos A, Berho M, Blumencranz PW, Brady MS. Giant cellular blue nevus of the anterior chest wall mimicking metastatic melanoma to the breast: A case report. *J Surg Oncol* 2000; 74: 278-281.
- Marano SR, Brooks RA, Spetzler RF, Rekate HL. Giant congenital cellular blue nevus of the scalp of a newborn with an underlying skull defect and invasion of the dura mater. *Neurosurgery* 1986; 18: 85-89.
- Elder D, Elenitsas R. Benign pigmented lesions and malignant melanoma. In: Elder D, Elenitsas R, Jaworsky C, Johnson Jr B, eds. *Lever's Histopathology of the Skin*. Eighth edition, Philadelphia:Lippincott-Raven 1997: 625-684.
- Moreno C, Requena L, Kutzner H, de la Cruz A, et al. Epithelioid blue nevus: a rare variant of blue nevus not always associated with the Carney complex. *J Cutan Pathol* 2000; 27: 218-223.
- Martinez-Barba E, Polo-Garcia LA, Ferri-Niguez B, Ruiz-Macia JA, et al. Congenital giant melanocytic nevus with pigmented epithelioid cells. *Am J Dermatopathol* 2002; 24(1): 30-35.
- Iemoto Y, Kondo Y. Congenital giant cellular blue nevus resulting in dystocia. *Arch Dermatol* 1984; 120: 798-799.
- Bennaceur S, Fraitag S, Teillac-Hamel D, Bodemer C, et al. Naevus bleu geant congenital de l'extremite cervico-cephalique. *Ann Dermatol Venereol* 1996; 123: 807-810.
- Mc Donagh AJG, Laing RW, Harrington CI, Griffiths RW. Giant alopecic nodule of the scalp: unusual presentation of a cellular blue naevus in an adult. *Br J Dermatol* 1992; 126: 375-377.

Blue nevi are typically benign tumors. However, although rare, malignant transformation of cellular blue nevi can occur. The risk of malignant transformation seems to be higher for giant form of the cellular blue nevi (3,12,13,18). The risk is higher for lesions on the head and in males (8). Giant cellular blue nevi have also a potential for local invasion of muscle, bone and meninges (8). For these reasons, giant cellular blue nevi should be considered potentially aggressive, regardless of their histologic appearance, and early total excision should be performed, particularly for the lesions located on the scalp (3).

Malignant transformation does not occur in the common blue nevi. However, in all of the reports about giant common blue nevi, no follow-up information was given. For this reason, it is difficult to assess the prognosis of giant common blue nevi, but the risk of malignant transformation seems to be very low. In our case, clinically, the lesion simulated malignancy because its large size and dark color. But, histologically the cells were devoid of mitoses, atypia, and infiltrative and destructive growth pattern as seen in malignant forms. There was no evidence of recurrence during the follow-up of 17 months.

In conclusion, congenital giant common blue nevi are very rare lesions. Because of their rarity and the lack of long-term follow-up in reported cases, it is difficult to assess the prognosis in terms of recurrence or malignant transformation. Although the risk of recurrence or malignant transformation seems to be very low, long-term follow-up is recommended.

10. Findler G, Hoffman HJ, Thomson HG, Becker L. Giant nevus of the scalp associated with intracranial pigmentation. J Neurosurg 1981; 54: 108-112.
11. Nakano S, Kinoshita K, Morita Y, Inoue S, et al. Cellular blue nevus of the scalp associated with intracranial involvement. Surg Neurol 1987; 27: 553-557.
12. Pathy AL, Helm TN, Elston D, Bergfeld WF, et al. Malignant melanoma arising in a blue nevus with features of pilar neurocristic hamartoma. J Cutan Pathol 1993; 20: 459-463.
13. Plantin P, Le Roy JP, Zagnoli A, Guillet G. Mélanome malin développé sur naevus congénital géant a type de naevus bleu. Ann Dermatol Venereol 1987; 114: 1243-1246.
14. Upshaw BY, Ghormley RK, Montgomery H. Extensive blue nevus of Jadasson-Tieche. Surgery 1947; 22: 761-765.
15. Pittman JL, Fisher BK. Plaque-like blue nevus. Arch Dermatol 1976; 112: 1127-1128.
16. Radentz WH, Vogel P. Congenital common blue nevus. Arch Dermatol 1990; 126: 124-125.
17. Kawasaki T, Tsuboi R, Ueki R, Fujioka N, et al. Congenital giant common blue nevus. J Am Acad Dermatol 1993; 28: 653-654.
18. Tuğyan N, Ermete M, Sarı AA, Doğan M. Malign blue nevus: İki olgu. Patol Bülten 2002; 19: 47-49.

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