# A neonate with malignant ectomesenchymoma

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**SUMMARY: Bayram İ, Leblebisatan G, Yıldızdaş H, Tuncer Ü, Ergin M, Narlı N, Tanyeli A. A neonate with malignant ectomesenchymoma. Turk J Pediatr 2005; 47: 382-384.**

**Malignant ectomesenchymoma is a rare tumor reported in head-neck, abdomen and perineal regions. It consists of mesenchymal and neuroectodermal elements. In this tumor group, neoplastic cells are differentiated into neuronal cells. It also has at least one malignant mesenchymal element, generally rhabdomyosarcoma. In this report we present a neonate with ectomesenchymoma.**

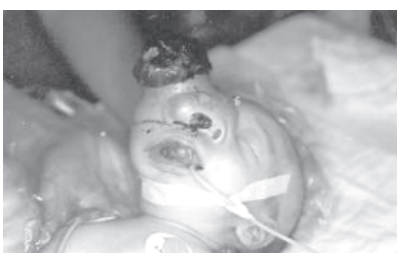
***Key words: malignant ectomesenchymoma, neonate.***

Malignant ectomesenchymoma is a rare tumor in childhood. It includes mesenchymal and neuroectodermal elements. It is a member of the biphenotypic tumor group and has been reported in the head-neck, abdomen and perineal regions. It is thought to originate from remnants of neural crest cells. Neoplastic cells are differentiated into neuronal cells in ectomesenchymoma. This tumor also includes at least one malignant mesenchymal element, mostly rhabdomyosarcoma[1-6].

Here we present a newborn infant with ectomesenchymoma.

### *Case Report*

A 10-day-old girl was admitted to hospital for evaluation of mass on her face (Figs. 1, 2). She was born to a healthy mother at 36 weeks of an uneventful pregnancy. Her weight was 3200 g (75-90 percentile) and length 52 cm (75-90 percentile). Vital signs were normal. Physical examination revealed a mass about 8x9 cm including mouth and nose extending to the hard palate with hemorrhagic and necrotic parts. Other system examinations were normal. Laboratory investigations showed normal blood count and biochemical values. The biopsy specimen was rich in reticulin and mitotic activity (6-8 mitoses in one high power field). ABC immunoperoxidase revealed desmin and SMA diffusely, and NSE and CD-34 were focally positive in the presence of signs of rhabdomyoblastic differentiation. Tumoral tissue was also examined for S-100 and keratin and showed positive reaction with mature ganglion cells. Based on the tumor's rhabdomyoblastic and neuronal elements, ectomesenchymoma was diagnosed (Fig. 3). No central nervous system involvement was detected by cerebral tomography. Because of mass localization, no surgical intervention could be done. Vincristine (1/2 of 1.5 mg/m$^2$), ifosfamide (1/2 of 1.8 g/m$^2$/day for 5 days with Mesna), and etoposide (1/2 of 100 mg/m$^2$/day for 5 days) infusions were started and adjusted



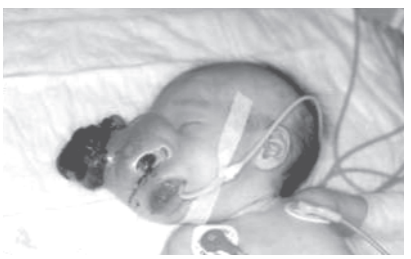


**Fig. 1, 2.** Anterior and lateral views of the case.

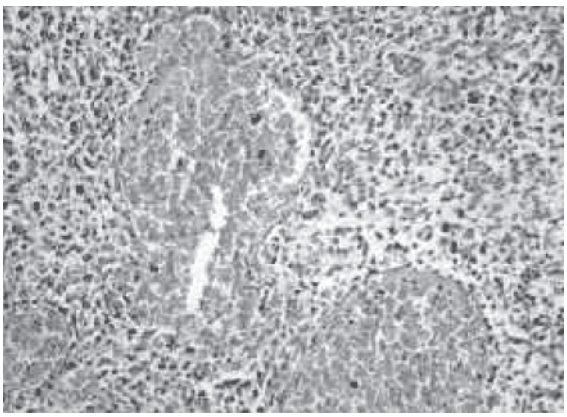


**Fig. 3.** Immature mesenchymal cells and vascular structures (HEx250).

according to the weight changes. The lesion was necrotic and bleeding. It continued to enlarge despite chemotherapy. Radiotherapy was planned but the case died due to sepsis as she had bone marrow depression.

## Discussion

To our knowledge there are about 60 case reports about malignant ectomesenchymoma. It has male preponderance and has been reported in all ages ranging from neonate to 60 years. About one third of all cases are under one year and two third are under seven years old[1,8]. The median age of appearance is 10 months. It has been shown by retrospective studies that most of the cases were initially misdiagnosed as rhabdomyosarcoma[8], so the rate of occurrence may be more than is reported.

Ectomesenchymoma is a highly malignant tumor. It spreads directly to surroundings: hematologically to lungs, liver, and intraperitoneal region, lymphatically to other tisses[1]. It is treated by a combination of chemotherapy, radiotherapy and surgery. If widespread metastasis and bone marrow involvement are present, cure is low[8].

In ectomesenchymoma, histological examinations show neuroectodermal and mesenchymal myogenic element[1,3-6,8]. Forty-nine percent of cases show ganglion cells, 25% ganglioneuroma, 12% neuroblastoma and 17% neural elements. In 82% of the cases, rhabdomyoma cells were reported[1,3-6]. In the present case, we detected rhabdomyoblastic and neuronal cells in the biopsy since specific markers were available. In electron microscopy, characteristic fibers of myofilaments, Z bands, rhabdomyoblasts, filaments, tubules, rough endoplasmic reticulum, nuclei and nucleoli and ganglion cells can be seen[9,10]. Unfortunately, we could not examine the biopsy by electron microscopy. The potential chromosomal defects were reported to be t(11-22), (q24-q12), FLT1/EWS and t(2-13)[9,11].

The treatment approach is wide surgical resection, chemotherapy and, if needed, radiotherapy. As a chemotherapy protocol, drugs used in rhabdomyosarcoma are used because of the high percentile of rhabdomyosarcoma component[1,3,4,6,7,12]. After achievement of remission, there is risk of relapse[6], so patients must be followed closely.

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