NEONATAL NEUROBLASTOMA

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ABSTRACT

Neuroblastoma is the most common malignant neonatal tumor but its presentation varies enormously. It may present with mass abdomen, lymphadenopathy or simply with diarrhea. A high level of clinical suspicion is required to diagnose the case. We present a case of neonatal neuroblastoma who presented with skin lesions. Diagnosis was established by biopsy of a representative skin lesion.

Key words: Neonatal Neuroblastoma, malignancy, metastases.

INTRODUCTION

Neuroblastoma is the most common malignant neonatal tumor, making up about 50% of neonatal malignant tumors[1] with an estimated incidence of 0.61 per 100000 live births[2]. The presentation varies, it may be massive or minute, irregular and stony hard. Rarely, it can present with diarrhea, hypertension[3], an enlarging, firm, usually painless, mass of the neck with Honer's syndrome[4,5] or even with cutaneous metastases[6], and exophthalmos[7]. Treatment varies from spontaneous regression to surgery and chemotherapy. We present a case of neonatal neuroblostoma who presented with nodular skin lesions of variable ages, diagnosed by biopsy of the representative skin lesion.

A 25 days old male child presented with multiple swellings on the body. According to the mother, the disease started with a single nodular swelling on the chest a few days ago. Since then, new lesion were appearing every second to third day and the disease had spread to involve the head, trunk and both the upper and lower limbs. There were no constitutional symptoms. Examination revealed an alert well looking neonate with multiple nodules of the whole body, which were of variable sizes. The location of the lesions was also variable. Some lesions were intradermal, some subcutaneous while others were deeply located. They involved the head including the left upper eyelid; the trunk and both the upper and the lower limbs. However, the trunk was relatively thickly populated. Both the thighs were swollen, with the left more than the right. Scrotum and prepuce were also swollen (Fig. 1 & 2).

The rest of the systemic examination was unremarkable.

Full blood count, urinalysis, X-Ray chest and skeletal survey showed normal results. Ultrasonography was unremarkable. Biopsy from a representative skin lesion (nodule) was taken. Microscopic picture showed tumor composed of groups of loosely packed small sized round to oval cells having fibrilla processes and interspersed with few gaglion cells.

The groups of cells were separated by fibro vascular septa and storma with evidence of rosette formation-findings consistent with metastatic neuroblastoma.

Further search for primary was not fruitful and the patient was referred to the Institute of Radiotherapy and Nuclear Medicine (IRNUM) for further management, with a request for follow up visits to our out-patient department.

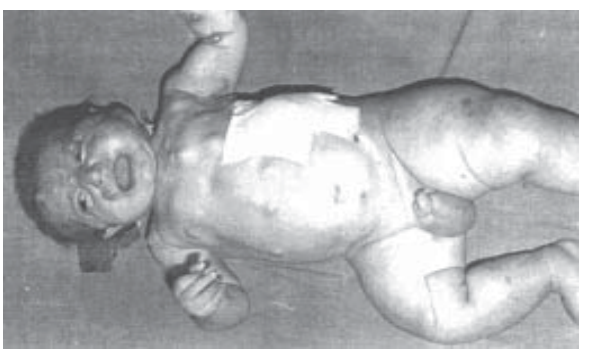

Fig. 1

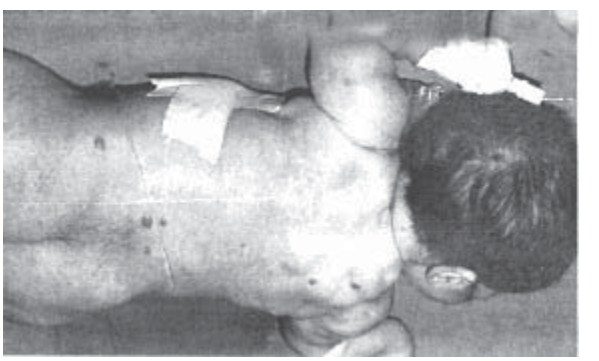

Fig. 2

The patient did not return to us for follow up and it was impossible for us to follow him at this village because he belonged to a far flung area of Mohmand Agency, a tribal area of N.W.F.P.

DISCUSSION

As a whole, neonatal malignancies are rare. They comprise 2% of all pediatric malignancies[2]. Neuroblastoma, the commonest neonatal malignancy, comprise of 50% of all neonatal tumors[1], the incidence being 0.61 per 100,000 live births[2]. The commonest presentation is with flank or abdominal mass, which is usually painless. However, the presentation may vary from simple diarrhea to as complex as "dancing eyes, dancing feet syndrome"[3,8]. Cautaneous metastases may be the only manifestation and hence of paramount importance[6] because the nature and the site of the likely neoplasm can be suggested by such surface clues[9]. Our patient presented with cutaneous metastases and we were unable to find out the primary due to the non-availability of computerized tomographic (CT) scanning and magnetic resonance imaging (MRI) facilities. The ultrasound scanning was not informative in this regard. The swelling of thighs, scrotum and prepuce suggests lymphatic obstruction due to compression effects.

Biopsy of the skin lesion rules out the commonest differential diagnosis i.e. nodular fat necrosis[10] and Torre's syndrome[6].

Treatment of the condition depends upon the site and the extent of disease.

A stage 4S disease may remit spontaneously[2]. Our patient seemed to have stage 4S because no evidence secondaries to any part of the body except the skin, was found, and hence our patient had a chance of good prognosis. A stage 4S disease is composed of a primary tumor stage 1 or 2, with spread limited to the liver, skin or bone marrow [2].

Prenatal diagnosis by ultrasound improves the prognosis[11] and survival may reach up to 90%[12]. Moppet J et al has an over all survival rate of 91%[2].

Initial bone marrow transplantation has given encouraging results[13].

CONCLUSIONS

The outlook for neuroblastoma is better than for any other tumour in the neonates.

The high risk of complications but good survival rates should be taken into account when counseling the parents.

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