NEUROBLASTOMA PRESENTING AS ECCHYMOSIS OF THE EYELIDS*

BY

D. J. WILSON

Manchester

WITH the exception of leukaemia, neuroblastoma sympatheticum is the most frequently occurring malignant neoplasm in infancy and childhood (Bodian and White, 1952). It is classified with neurological tumours, because it originates from tissue which gives rise to the adrenal medulla and portions of the sympathetic system. The most common single site is the adrenal medulla (Willis, 1960), but it can arise from sympathetic tissue anywhere in the body.

The main histological features are numerous cells, like lymphocytes, with a scanty stroma, rosette formation, and neurofibrils.

Metastasis takes place by both lymphatic and, especially, blood stream invasion.

Hutchison (1907) drew attention to his "clinical syndrome", describing seven cases of tumour of one or other adrenal with metastases in the bones of the skull. In two of his cases ecchymosis of the eyelids was the first physical sign. Bothman and Blankstein (1942), discussing the importance of ocular findings in neuroblastoma, reported four cases observed by them in 1936-37. *In one of these, ecchymosis of an eyelid was the presenting sign. Despite this, it is not generally realized that neuroblastoma not infrequently presents as ecchymosis of the eyelids, as in the following case.

Case Report

A female infant aged 10 months was brought by her parents to the Out-patients Department of the Royal Eye Hospital, Manchester, in February, 1959. It had been noticed 5 weeks previously that the skin was black under the infant's left lower eyelid. This was followed by a sub-conjunctival haemorrhage on the left side. The bruising increased in size for 2 weeks and then receded, but it had recurred, and for 2 weeks before the child's attendance at the hospital the eye had been obviously proptosed. The child was also suffering from a "cold". There was no history of injury. She was very ill and was admitted to hospital for investigation.

Examination.—On admission the temperature was 104.6°F., and the pulse and respiration rates were 145 and 28 per minute respectively. No abnormalities were found in the cardiovascular or respiratory systems. The liver was greatly enlarged and a nodule was palpable at its edge.

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The blood count showed haemoglobin 7.7 g. per 100 ml. (52 per cent.), red blood cells 3.46 million per cu. mm, colour index 0.76, white blood cells 25,800 per cu. mm.

The differential count showed polymorphs 40 per cent., lymphocytes 56 per cent., mononuclears 4 per cent. Aniso- and poikilocytosis were rather marked and polychromasia was slight. The platelet count was not done.

The patient's condition rapidly deteriorated and she died within 48 hours of admission.

Post mortem Examination.—A large tumour mass was found involving the right adrenal and right kidney. Secondary deposits were found in the liver, pancreas, and ribs. In the skull the ethmoid was largely replaced by a mass of haemorrhagic tumour which was soft and had eroded the bone. It had pushed itself up anterior to the pituitary fossa and extended laterally to include both optic nerves. On the left the tumour had extended across, or round, the cavernous sinus to the middle fossa and had invaded the lateral aspect of the left orbit. Although the tumour surrounded the optic nerve it had not invaded the eye. The histology of the tumour in the liver (Figure) was characteristic.

FIGURE.—Section of secondary deposit of the neuroblastoma in the liver, showing typical arrangement of neuroblastoma cells with intervening fibrillary zones. $\times 180$.

Discussion

In view of the history of ecchymosis as the first sign in this case, the records of the Manchester area were examined. It was found that 36 cases of neuroblastoma sympatheticum were included in the Children's Tumour Register of the University of Manchester between September, 1953, and

September, 1958. Of these six had presented with ecchymosis of the eyelids as the first sign, and in one of these cases bruising was the only sign for 4 weeks (Steward, 1960).

Leukaemia is the other important disease producing ecchymosis of the eyelids in childhood. Examination of blood and bone marrow should leave little doubt as to the correct diagnosis, for the haemorrhage in leukaemia, which results from a fall in the platelet count, occurs only when the white blood cell count is abnormal. Haemorrhage in neuroblastoma, on the other hand, is due to the local effects of the secondary tumour deposit and the blood picture is not characteristic. There may be an accompanying secondary anaemia, but the distribution of white blood cells in the peripheral blood is normal. The interpretation of the bone marrow findings in neuroblastoma is not easy.

Summary

A case of neuroblastoma presenting as ecchymosis of the left lower eyelid and conjunctiva is reported. An attempt has been made to show that bruising of the eyelids is not an uncommon presenting sign in neuroblastoma and may, therefore, be of some importance as an aid to early diagnosis. Leukaemia may also present as bruising of the eyelids. The bone marrow findings and particularly the blood picture are of basic importance in differentiating one disease from the other.

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NOTES

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