HÆMORRHAGIC THROMBOCYTHÆMIA

A CAUSE OF REPEATED SEVERE EPISTAXIS

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THE purpose of this article is to underline a cause of repeated epistaxis which is rarely recognized, but which may be more frequently diagnosed as rhinologists become increasingly aware of hæmatological possibilities.

Thrombocythæmia is a rare disorder in which patients bleed spontaneously from mucous membranes, particularly from the gastro-intestinal tract and they also show a tendency to excessive bleeding after minor surgical operations such as dental extraction.

It has been defined as a proliferative disorder of the megakaryocytes of the bone marrow resulting in an extreme peripheral thrombocytosis (Stefanini and Dameshek, 1955).

The example quoted later demonstrates the ease of diagnosis with adequate hæmatological investigation and the possibilities of relief of symptoms, and avoidance of more major catastrophe.

The name for the condition was first suggested by Epstein and Goedel in 1934 but cases in the literature have been variously named as thrombocytosis, idiopathic or primary thrombocythæmia. More recently there has been a tendency among hæmatologists to group it with the myeloproliferative disorders such as granulocytic leukæmia and polycythæmia vera. Gunz (1960) defines the condition as "a clinical syndrome with presenting symptoms of recurrent spontaneous hæmorrhages, either external or into the tissues, and often preceded or accompanied by thromboses in the superficial and deep veins. There is splenomegaly in the majority of cases. The blood picture shows an extremely high platelet count and usually a high neutrophil leucocytosis". Hypochromic anæmia is usually present.

About fifty cases are reported in the literature. Males and females appear to be equally involved and the average age is 51. Gastro-intestinal hæmorrhage is most frequently mentioned but bleeding after minor trauma and operations, menorrhagia, hæmaturia, hæmoptysis, epistaxis and excessive bruising are also described.

Autopsy findings are poorly described in the literature, but the out-

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standing findings are bone marrow hyperplasia, particularly affecting the megakaryocytes, usually with excessive platelets. There is splenomegaly, from slight to gross, in nearly all cases, and hepatomegaly occasionally with myeloid metaplasia. Thrombotic lesions of blood-vessels are common, especially in the splenic vein. The causes of death are usually uncontrollable hæmorrhages or thrombo-embolic lesions of gastro-intestinal vessels or cerebral circulation.

The platelet count varies from 900,000 to 14 millions per c.mm. with an average of 3·2 millions per c.mm. The platelet shapes are very variable and bone marrow shows excessive platelet agglutinates. Most authorities agree that the degree, frequency and amount of hæmorrhage is related to the platelet count. About half the reported cases showed abnormal bleeding times (Fountain, 1958). Excessive bleeding in the presence of excessive platelets appears at first sight to be paradoxical, and the reason for the excessive hæmorrhagic tendencies is not understood. Benney and Lewis suggest the possibility of a defective Christmas factor as the cause of the excessive bleeding (Benney and Lewis, 1959).

Most patients suitably treated appear to have an expectation of life of at least several years, but splenectomy for splenomegaly converts a chronic disorder into an acute illness. The normal progress of the condition appears to be a slow one but untreated cases tend to bleed spasmodically and later thrombotic episodes may lead to cerebral catastrophe or gastro-intestinal hæmorrhage from infarction.

Therapy is directed towards stopping megakaryocytic proliferation by the administration of radioactive phosphorus (P. 32) (5-10 millicuries can be given in divided doses orally or intravenously). This therapy can be repeated at intervals until the platelet count falls to normal, usually in three or four weeks.

Triethylene melamine (TEM) has also been used but has considerable side effects. Urethane and X-ray therapy to the skeleton have also been used.

Blood transfusion is required in severe hæmorrhage, and where thrombosis has occurred heparinization may be indicated to stop extension of the thrombus.

R.L., a male, aged 73, was admitted under my care to the General Infirmary at Leeds. He had suffered from slight recurrent epistaxis for 20-30 years, but during the five months prior to admission he had three bouts of severe epistaxis, involving both sides, and severe enough to require packing on several occasions. There was no known precipitating factor such as trauma or infection and when first seen, apart from blood clot in the nose, there was no abnormality in nose or nasopharynx. He admitted to bruising easily, had ankle œdema and varicose veins, and had had angina of effort for a year. Sinus X-rays were normal. B.P. was 175/90.

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Hæmatology

7.4.61 Hæmoglobin: 68 per cent.
Bleeding time: more than 10 minutes.
Clotting time: 9 minutes.
W.B.C.: 30,000 per c.mm.
9.4.61 Platelets: 2,660,000 per c.mm.

Repeated platelet count over the next few days showed a count of between one and two millions per c.mm.

Sternal marrow puncture on 18.4.61 was reported as follows:

"The prominent feature of the marrow is the large numbers of the megakaryocytes at all stages of maturation. This megakaryocytic hyperplasia is too pronounced to be accounted for by bleeding alone, and would be in keeping with a diagnosis of hæmorrhagic thrombocythæmia. Erythroid hyperplasia is a prominent feature; the late normoblasts, however, are well hæmoglobinized."

He was given 3 millicuries of radioactive phosphorus intravenously with the platelet level at 1,420,000. Three weeks later he was feeling well and had no further epistaxis, but his platelet count was still 1,360,000. The dosage of P.32 was, therefore, repeated, and one month following this his platelets had fallen to 420,000 per c.mm., his hæmoglobin was 92 per cent. and white cells 7,200 per c.mm. He said he was very well and had no further bleeding. His condition will be under review.

Conclusions

Thrombocythæmia as a cause of repeated severe epistaxis is described.

It is probably closely related to other myeloproliferative disorders. Its ætiology is unknown, as is the relationship of excessive platelets to excessive bleeding. All rhinologists are fairly frequently confronted with severe repeated epistaxis, the cause of which is obscure, and occasionally with fatal termination. In such cases the diagnosis of thrombocythæmia is easily missed unless a platelet count is asked for specifically. In cases of doubt the diagnosis can be confirmed by sternal marrow puncture. The treatment by P.32 appears to be efficient.

It would seem that repeated nasal packing with gauze strips probably aggravates the epistaxis due to the trauma necessarily incurred, and where pressure on the bleeding area is really necessary, the use of a Foley catheter might be less traumatic. Thrombocythæmia may be a more common cause for epistaxis in the adult than is at present appreciated.

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