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LETTER TO THE EDITOR

YOUNG STROKE DUE TO F XIII DEFFICIENCY

Sir,

(Received on May 29, 1997)

The discovery of F XIII by Laki and Lorand (1) also known as Fibrin Stabilising Factor is an important landmark in the field of coagulation physiology. This is a plasma protein which fibrin. Defficiency of this factor also cause haemorrhagic state (2). Ducket et al (3) were the first to observe a case of haemorrhagic disorder due to congenital defficiency of F XIII. Since then more than a hundred cases have been reported in Medical literature, however, such a case was first to be observed by us.

Patient summary

SD, a 18 yrs old female got admitted in a stuporous condition with left sided complete opthalmoplegia and right sided hemiplegia of 3 days duration. There was no past history of bleeding from any site, drug intake or menorrhagia, but her father recalled delayed would healing since early childhood. Family history was strongly suggestive, out of 8 sibs, the first died at the age of 2 yrs with h/o massive bleeding from umblical site that required frequent transfusion, the second died at the age of 9 months from G.I. bleed, the third was a neonatal death with massive haematomas under the skin, the forth died of massive bleed from nose and mouth at the age of five years and succumbed. The fifth child is the case under discussion. The sixth has h/o bleeding from ubmlical site, doing well, found to be F XIII defficient. The seventh sib has not shown any bleeding diathesis, F XIII defficient so also the eighth.

Clinical Examination showed features of left sided complete opthalmoplegia and right sided hemiplegia. There was papilloedema both sides, drowsy state, cardiac evaluation normal, peripheral arterial pulses normal, no bleeding from any other site. On investigation found to have mild normocytic normochromic anaemia (Hb–10.4 gm%), Platelet count: 2.3 lakhs/cmm, Reticulocytes – 3%, TLC–11, 900/cmm, DC–N:74%, L:19%, M:4%, E:3%. The coagulation parameters revealed BT – 3 min (Normal 2–5 min), Prothrombin Time–12 sec (Normal 11–14 sec), Prothrombin Consumption Index–45% against a normal of 0–40%, APTT–43 sec against a normal value of 35–45 sec, Fibrinogen–320 mg against a normal of 200–400 mg, Russel's Viper Venom Time–16 sec against a normal of 15 sec, Factor Assay for II, V, VII, VIII, IX and X were all normal. Clot Solubility test, strongly positive. The study of F XIII was done by

Results of the solubility tests of the fibrin clot.

Sample from	*Solubility of the clot*	
	5 M Urea	*1% Monochloroacetic acid*
Control	4+	2+
Patients	0	0
Father	4+	2+
Mother	4+	2+

observing the solubility of a fibrin clot obtained by recalcification of Oxalated plasma. The results were graded from 0–4+ according to the size of the remaining clot.

The abnormal clot solubility test and slightly raised prothrombin consumption Index indicated isolated F XIII defficiency. The CT scan of brain showed a dense high attenuated area seen in left temporo–parietal region without any mass effect with Housefield unit of 60.5 corresponding to that of blood, size of haematoma being approx 3.7 × 2.9 cm.

The patient was managed with conservative treatment, anti oedema measures, 4 units of blood transfusion, after 10 days staying at the hospital she was discharged followed up for 6 months, doing well.

DISCUSSION

The number of affected siblings of both sexes in this family studied shows that the diseases of F XIII defficiency is hereditary, autosomal transmission (4). The characteristic feature of bleeding in the family was from umblical site, rarely reported in literature (5). Intracerebral haemorrhage very rarely seen in other coagulation factor defficiency states is remarkably frequent in F XIII defficiency (6). Since the patient improved dramatically with fresh blood transfusion, the search for isolated F XIII supplementation was not thought of.

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