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REPORT OF THE CASE OF IDIOPATHIC MYOGLOBINUREA (MEYER-BETZ DISEASE)

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Meyer Betz was the first who reported a case of Idiopathic Myoglobulinaemia in 1910[1], for this reason the disease called Meyer Betz disease. The involved Children presents with paraxusmal myalgia accompanied with changes in the color of the urine.

In this article we are presenting a case history with a short discussion, about the etiology, pathophysiology and metabolic changes of this disorder.

PRESENTATION OF THE CASE:

A 3 year-old boy, living in Tehran was admitted to the hospital with a chief complain of inability to walk. He was doing well till 5 days prior to admission, when he had generalized weakness, malaise associated with anorexia and vomiting which lasted for two days. He showed no improvement by the usual treatment, and referred to this hospital. In the second day of his illness, his urine became dark-brown in color which was lasted for 48 hours, when the color restored back to its normal color.

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In his past medical history, the boy had Measles whooping cough at age two. Epistaxis, one month before his present illness due to trauma to his face. He is the forth child to the healthy related-cousin-family. Two sisters eight and twelve yeas old and one brother 6 yeas old.

On examination the patient was in sick looking afebrile. His head and neck appeared normal. Mucous membrane and conjuctivita were not anemic. Light reflex was normal. He was slowly talking without evidence of paralysis of the uvula, and his lymphnodes were not enlarged.His pulse rate was 160 per minutes with normal heart sounds without murmur.

GASTRO INTESTINAL EXAMINATION:

Patient was nauseatic and showing difficulty in swallowing solid food but not for fluid. He had normal motion. His liver and spleen were not enlarged and his Genitourinary Tract were normal.

The posterior calf muscle was tender, firm and painful. Also the muscles of the thigh were painful. He was unable to move without help. The power of the muscle was decreased in both legs with flacid paraplagia. His reflexes and sensation were normal, so his cranial nerves.

The urine was normal in color. Examination of the sediment was negative. The stool was normal and showed no parasites or undigested food. The sedimentation rate was 8 in first hour and 22 in second hour. The hematocrit was 34%, the Hemoglobin was 9.6 gm/100ml,Platelets count were normal. His WBC was 9600, with 56% segmented form, 36% lymphocytes, 4% monocytes and 4% Eosinophils. Cerebro-Spinal Fluid was within normal limit. An X-ray

of the chest showed increased Pulmonary Vascular marking in the hilum with slight haziness in bibasalar area of the lungs.

On the 22nd days the patient discharged from the hospital in good general condition with restoring his ability to walk without help.

15 months later he was re-admitted with fever, in ability to walk, anorexia, coughing and chest pain.The sickness was of 2 days duration prior to admission and his urine color changed to dark brown since then. On examination, weight was 12.5 Kgms. Head and neck were normal. He was conscious and answering the questions well. He had no difficulty in swallowing and light reflex was normal. He was tachycardic with heart rate of 148 per minute. Bp $\frac{120}{70}$. Lungs were clear. Liver and spleen were not enlarged, and had no lymphadenopathy. His Gentourinary tract were normal and his urine was dark brown color. The legs muscle were tender and painful, and he kept both lower limbs in extension position. He was unable to sit and the passive movement of the legs was painful. His lower limb was tender, reflexes were sluggish, but planter reflex was normal and so his sensation test. The tone of the upper limb muscles were decreased. Cranial nerves were normal.

In the second day of admission the urine color turned to normal. He was oliguric for 2 weeks with daily urine output of 100-150 cm., and during this period his urine color turned twice to dark brown color, and gave a positive test for protein with 2.7-3 gm,,albumin/lt and 1-2 WBC and RBC power field. Mir reaction was weakly positive. Afterward in the post oliguric phase, the urine was nega-

after heavy effort in adults mainly in solders (Half disease).

The etiology is unknown, although an abnormal metabolism in the muscles which is congenital in origin was suggested. Exercise, prolonged movement or infection is a stimulator to this abnormal metabolism, which leads to lysis and damage to the affected muscles. Most of the studies werc done on the muscle protein which is prodominently of fetal protein type in this disorder.[5-6] In the timebeing, Perkoff theory is more accepted, said that the disease is due to the presence of abnormal protein (Fetal protein) in the muscle cells. Although the study of Hed in 1955 showed that the main problem is in the Carbohydrates of the muscle cells[7]. Clinically this was suggestive, for a high calories requirement, like during infection or after effort, decreases the frequency of the attacks.

The observation of Heinz in 1965 revealed increase in lactic acid production during the episodes,this postulates that the basic disturbance is in the carbohydrate metabolism and mainly involving Krebs cycle, or probably it is a congenital disorder affecting the glycogen of the muscle cells with increase in the activity of pros phorelate enzymes[8].

As reported in most studies, the disease is presented with muscular pain, weakness proceded by fever, walking and movement abnormality, however, leukocytosis and fever are more frequent in the second type. The brown color of the urine is due to presence of myoglobin in the urine, which is the leading cause of death in this disorder, ending with reveal failure or tubular necrosis.

Although is some cases death was due to hyper Kalemea and sudden death.

The abstract we reported was for a 3 year boy with clinical signs and symptems of Meyer-Betz disease or myoglobinuria. A summary of the metabolic abnormality were mentioned and discussed briefly.

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La localisation du lymphome malin du SNC est habituellement dans le cerveau et plus rarement dans la moelle epiniere.

Nous rapportons ici un cas de lymphome malin cerebral avec etude histologique et tomodensitometrique.

Materiel et Methode

Donnees cliliques

Notre malade est un homme de 60 ans ayant presente neuf mois avant l'hospitalisation c'est-a-dire au debut de 1984, une hémiparésie droite accompagnée de troubles de conscience et perte de memoire.

Le patient est admis dans une clinique de Teheran.A l'examen on constate une parésie importante de l'hémicorps droit, des troubles de conscience, une apathie et une obnubilation. La numeration globulaire et la formule sanguine sont normales. Il n y a pas de protéinurie ni glycosurie. La tension artérielle est de 130/75 millimetres de mercure. La radiographie pulmonaire est normale.

Données Tomodensitométriques:

L'examen a été effectué par l'appareil scanner EMI 5005. Les coupes de 10mm. d'épaisseur ont été réalisées dans des plans paralléles au plan de virchow. 60 ml.de produit de contrate iodé ont été introduits en injection intraveineuse. La tome-densitométrie met en évidence,un changement archit-ectural tissulaire, au niveau du lobe frontal gauche avec une densité anormale qui possède une grande capacité d'absorption du produit de contraste.

Le phenomene d'absorption de cette néoformation est non homogéne, accompagné d'un oedeme périfocal notable. Le ventricule homolatéral est également collabé et les

structures des parties centrales ainsi que le système ventriculaire sont repoussés au côté contro-latéral. La loge postérieure est normale. Fig 1 et 2. En conclusion, la tomoden-sitométrie met en évidence une volumineuse tuméfaction néoplasique dans le lobe frontal gauche dont la morphologie est du type intra-cérébral. Un processus metastatique ou un méningiome est suspecté. Pour la verification histologique une biopsie cérébrale fut pratiquée.

Données anatomo-pathologiques:

Le materiel de prélévem-ent biopsique a été fixé au formol de 10% pendant 48 heures. L'inclusion a été faite en paraffine et les méthodes de coloration utilisées sont l'hémateine-éosine(HE),L'acide périodique de schiff(PAS) et réticuline. Histologiquement, le tissue cérébral est largement infiltré par des cellules lymphoides (fig 3). avec un grand noyau relativement clair (fig.4).Cette infiltration est soit diffuse, Soit fréquemment en manchons périvasculaires. Les cellules néoplasiques se localisent à l'interieure de la paroi et à la périphérie des vaisceaux. La Trame réticulinique est riche surtout autour des parois vasculaires. Fig 5 D'une maniere générale les cellules lymphoides infiltrent diffusément le parenchyme cérébral et ont tendance d'envahir particulierement les parois et les régions périvasculaires, formant des manchons autour des vaisceaux.

Evolution et Traitement:

A la recherche d'autres foyers dans l'organisme,une serie d'examens complémentaires fut pratiquée, Parmis lesquels nous citons notamment, La lymphangio-graphie,la biopsie osseuse de la crête iliaque et la scannographie du foie, sont normaux.

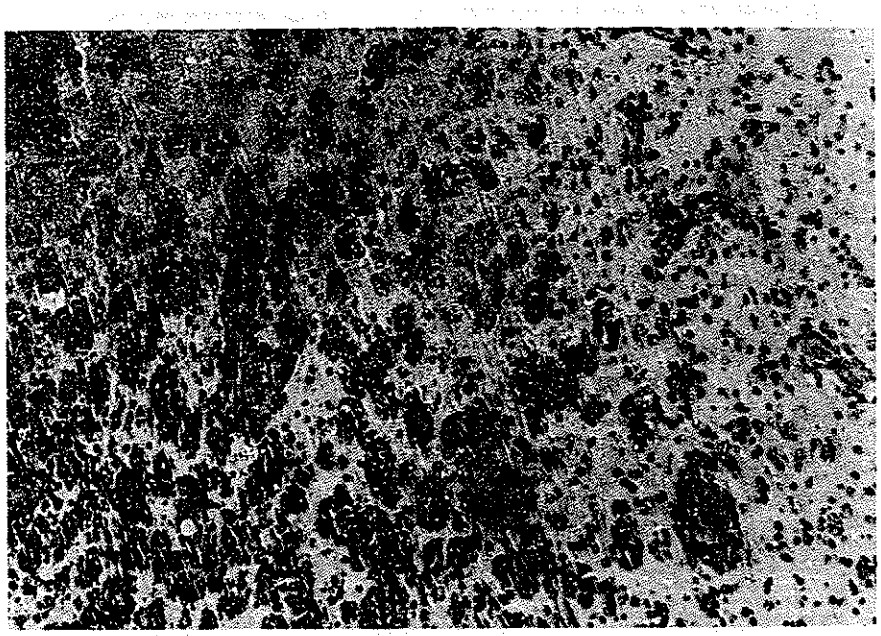

Fig III: Infiltration diffuse de cellules lymphoides. Paraffine-HE

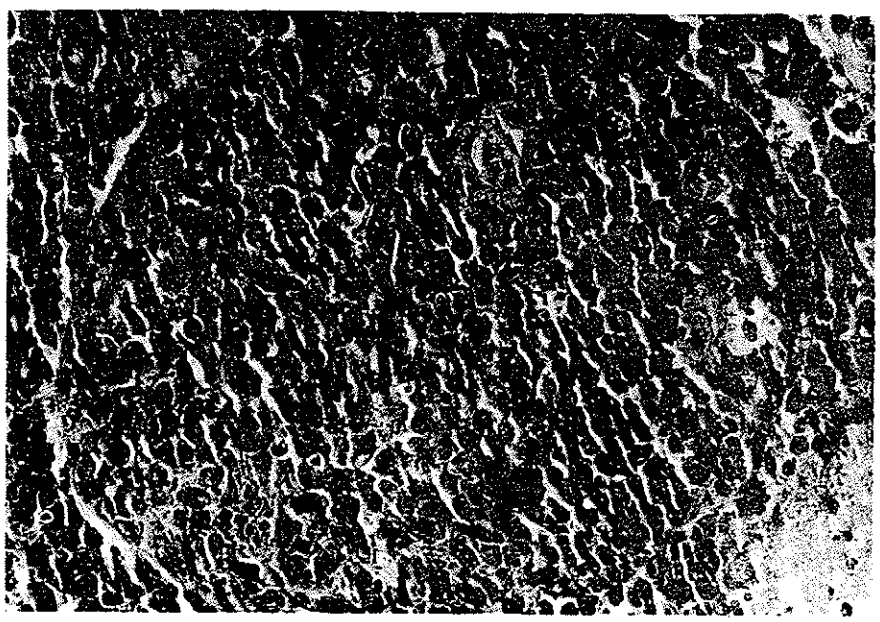

Fig.IV: Aspect de cellules tumorales. Noter que les cellules ont un noyau relativement grand et clair(paraffine-HE)

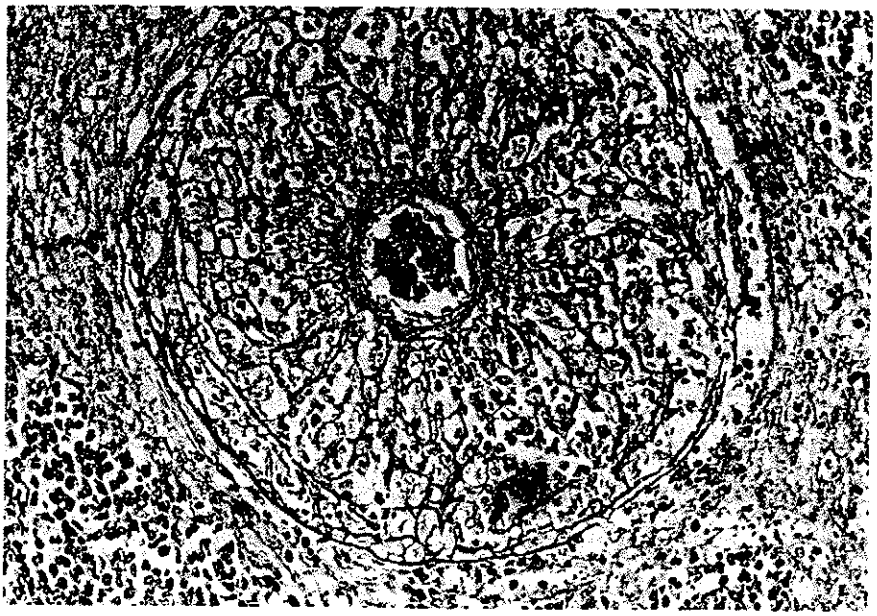

Fig. V: Infiltration tumorale en manchon péri-vasculaire. paraffine-Réticuline.

Discussion:

Depuis les premières descriptions, de nombreux exemples de lymphome malin du SNC ont été rapportés, aboutissant à des series importantes Permettant ainsi de dresser un bilan assez précis concernant les manifestations anatomo-cliniques et de discuter l'origine des cellules néoplasiques.

I. Caracteres cliniques et pronostiques

Principalement, les adultes sont atteints. L'age moyen est de 57 ans Les signes cliniques predominants chez un

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