

by reports from other regions of India(4-7) that the incidence of reticuloendothelial malignancies is highest in distribution of childhood cancer, and in our country we have a pattern of increasing trend of leukemia, medium glioma and medium lymphoma(7).

We conclude, that because of smaller number of children presenting with childhood malignancies at Medical Colleges, development of Regional Pediatric Oncology Centres are needed.

REFERENCES

1. Venugopal KV, Joseph TP, Verma KK. Solid malignant tumors of infancy and childhood. A clinicopathological study. *Indian Pediatr* 1981, 18: 365-368.
2. Gandhi RK. Cancer in childhood, need for planned approach. *Indian Pediatr* 1981, 18: 363-364.
3. US Department of Health Education and Welfare. Vital statistics of United States. Volume II. Mortality, Deaths from Selected Cause, 1964.
4. Nair NS, Nair Raman R, Mathew O. Neoplasms in children. *Indian Pediatr* 1964, 1: 26-32.
5. Paul SS, Shagurin O, Modi S, Kabra S, Singh G. Malignant neoplasm in childhood. *Indian Pediatr* 1967; 4: 309-313.
6. Thaper RK, Gupta MC, Verma JK. *et al.* Malignancy in childhood. *Arch Child Hlth* 1971, 13: 57-68.
7. Pathak IC, Datta BN, Aikat RK, Reddy MM, Bhattacharjee N. Pattern of neoplastic diseases in children with special reference to malignant tumors. *Indian J Cancer* 1975, 12: 46-55.

Cerebral Cavernous Angioma

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Cerebral cavernous angiomas are rare lesions. Approximately 350 cases have been reported in the literature and of these only 70 cases (20.6%) were eighteen years old or younger(1-3). Because of its rarity in children, the present case is being reported.

Case Report

A 14-year-old female child was admitted with complaints of recurrent episodes of diffuse, severe headache alongwith vomiting for two and a half years duration. These episodes used to occur at an interval of about 2 months and would last for about one and a half hours. There was no history of associated loss of consciousness or visual disturbances or tinnitus or parasthesias.

On examination, the higher mental functions, cranial nerves, upper limbs and lower limbs were normal. On investigating, hemogram and urinalysis were normal. ESR was 5 mm in 1st hour. Plain X-ray skull was normal. CT scan showed a high

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attenuating lesion in the left temporal region which showed no contrast enhancement (Fig. 1). There was no perifocal edema.

Left temporal craniotomy revealed a well defined encapsulated 1 cm sized bluish mass with small vessels over it. Histopathologic examination revealed the features of cavernous angioma in the form of dilated vascular spaces of varying size with flattened endothelium (Fig. 2). Postoperatively she is symptom free for the last one and a half years.

Discussion

Intracranial cavernous angiomas are congenital vascular malformations composed of thin-walled sinusoidal spaces

lined by endothelium. They lack in elastic and muscular layers and the intervening nervous tissue which differentiates them from telangiectasis.

Cavernous angiomas can occur at any age and are rare in children. On reviewing the literature, Gangemi *et al.*(3) found that youngest patient with this type of lesion, was of 2 days age. Largest series of 5 cases in children was reported by Pozzati *et al.*(4). Rarely it has been reported to be familial(5,6). The lesion may vary in size from petechiae to a nodule(6,7). Rarely they may appear as a cystic tumor or may show calcification(6-8).

Nearly 80% of the lesions are supratentorial and 20% infratentorial. Occasionally they may be present in subarachnoid cisterns as an extra-axial mass(9). According

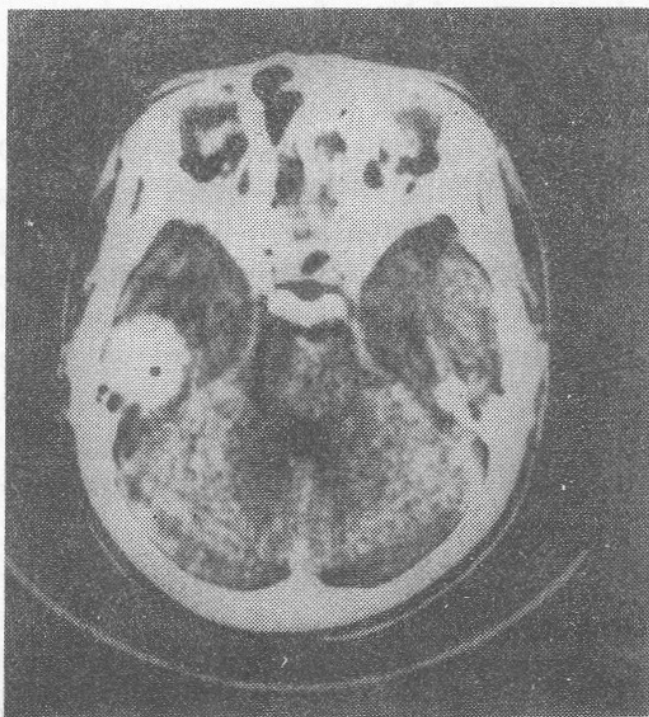


Fig. 1. Plain computerized tomographic scan showing a well-circumscribed roundish lesion of slightly uneven increased density in the left temporal region. After contrast injection there was no evidence of enhancement.

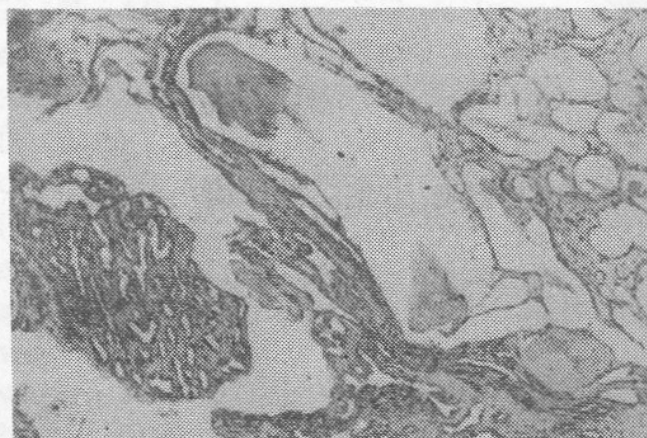


Fig. 2. Microphotograph showing vascular spaces of varying size, some of them being cavernous and lined by flattened endothelial cells.

to Giombini and Morello(5), the commonest presentation is seizure (38%), which may be generalized or focal. They may present as intracranial hemorrhage (23%), headache (28%) and focal neurological disturbance (12%). Our patient presented with the second commonest symptom.

As there are no feeding arteries and draining veins are usually small(6,7), so on angiography these lesions fail to opacify, hence they are also known as cryptic malformations. On angiography, cavernous angioma usually present as a non-specific vascular lesion but in some cases the angiography may be completely normal or in some there may be widened veins, a faint capillary blush and slight neovascularity(2). The recent advent of computed tomography has improved the recognition of these angiographically non-demonstrable angiomas. CT usually shows a roundish well-defined lesion with a slightly uneven high density often in the calcification range with minimal or no enhancement with intravenous contrast. Surrounding edema and mass effect is usually absent(10,11). Our case also showed high density lesion without any associated edema and did enhance with contrast. CT signs are nonspecific and have been interpreted as meningiomas, granulomas or ruptured arteriovenous malformation(12). With passage of time, angiomas may undergo fibrous and hyaline thickening of the vessel wall, intraluminal thrombosis and calcification(13).

Surgery is indicated to relieve the epilepsy and risk of intracranial bleed is taken care of. These lesions are generally well-demarcated from surrounding brain and can be resected without major neurological deficit. In cases with hemorrhage, it is important to obtain the tissue from the area surrounding it for histopathologic studies. We suggest that surgical excision of the ac-

cessible lesions is indicated because there is possibility of bleeding and the presence of a tumor can not be completely ruled out as in our case.

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REFERENCES

1. Voigt K, Yasargil MG. Cerebral cavernous hemangiomas or cavernomas. *Neurochirurgia* 1976, 19: 59-68.
2. Simard JM, Garcia-Bengochea F, Ballinger WE Jr, Mickle JP. Cavernous Angioma: A review of 126 collected and 12 new clinical cases. *Neurosurgery* 1986, 18: 162-172.
3. Gangemi M, Longatti P, Maiuri F, Cinalli G, Carteri A. Cerebral cavernous angiomas in the first year of life. *Neurosurgery* 1989, 25: 465-469.
4. Pozzati E, Padovani R, Morrone B, Finizio F, Gaist G. Cerebral cavernous angiomas in children. *J Neurosurg* 1980, 53: 826-832.
5. Giombini S, Morello G. Cavernous angiomas of the brain. Account of fourteen personal cases and review of the literature. *Acta Neurochir* 1978, 40: 61-82.
6. Russel DS, Rubinstein LJ. Tumors and hamartomas of the blood vessels. *In: Pathology of Tumors of the Nervous System*, 4th edn. 1977, pp 116-145.
7. McCormick WF. The pathology of vascular (arteriovenous) malformations. *J Neurosurg* 1966, 24: 807-816.
8. Ramina R, Ingunza W, Vonofakos D. Cystic cerebral cavernous angioma with dense calcification. *J Neurosurg* 1980, 52: 259-262.
9. Kawai K, Fakui M, Tanaka A, Kuramoto

- S, Kitamura K. Extracerebral cavernous hemangioma of the middle fossa. *Surg Neurol* 1978, 9: 19-25.
10. Savoirdo M, Strada L, Passerni A. Intracranial cavernous hemangiomas, neuroradiologic review of 36 operated cases. *Am J Neuroradiol* 1983, 4: 945-950.
 11. Terao H, Hori T, Matsutami M, Okeda R. Detection of cryptic vascular malformations by computerized tomography. Report of two cases. *J Neurosurg* 1976, 51: 546-551.
 12. Tagle P, Huete I, Mandez J, Del Villar S. Intracranial cavernous angioma: Presentation and management. *J Neurosurg* 1986, 64: 720-723.
 13. DiTullio MV Jr, Stern WE. Hemangioma calcificans. Case report of an intraparenchymatous calcified vascular hematoma with epileptogenic potential. *J Neurosurg* 1979, 50: 110-114.

Primary Generalised Lymphatic Dysplasia

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The manifestations of primary lymphatic dysplasia in children occur as a consequence of congenital maldevelopment of the lymphatic systems. Often, only a single site is involved and the presentation is with lymphedema or chylous ascites or chylothorax. The rare combination of all 3 components in a single patient is termed as generalized primary lymphatic dysplasia

and was first reported by McKendry *et al.*(1). Since then another eight authenticated case reports have appeared in the world literature(2,3). The rarity of the condition prompts us to report one such infant who, in addition to the other features, had a large chylopericardium.

Case Report

A 3-month-old boy, the first born in a non-consanguineous marriage, presented with gradually increasing abdominal distension since birth and fever for 2 days. Examination showed a febrile, anemic infant weighing 4 kg with tachypnea and bilateral pulmonary rales. In addition, he had non-pitting edema of the dorsum of the left hand and a soft, non-transilluminant swelling of the right side of the neck. There was a tense ascites and bilateral reducible inguinal herniae (*Fig. 1*). Investigations revealed a hemoglobin of 6.2 g/dl and polymorphonuclear leucocytosis, with an absolute lymphocyte count of $1.7 \times 10^9/L$. *Staphylococcus aureus* was isolated from the blood culture. An X-ray of the chest showed normal cardiac size, bilateral bronchopneumonia and mild bilateral pleural effusions. The total serum protein was 5.2 g/dl with an albumin fraction of 2.0 g/dl. Thoracic and abdominal paracenteses drew acellular, sterile milky fluid. This fluid had a specific gravity of 1015, fluid:

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