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Rare case of inflammatory pseudo tumor of central nervous system: A case report

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Inflammatory Pseudo Tumors (IPT) are mostly benign lesions, mimicking malignant lesions and affecting almost all organ systems and are characterized by fibrotic ground tissue and polyclonal mononuclear infiltrate on histopathology. It is a disease of unknown pathogenesis and the brain is a rare site of occurrence. Here we present one such case we operated recently in our department.

Introduction: Inflammatory Pseudo Tumor (IPT) is a rare nonneoplastic condition that usually involves the lung and orbit but is known to affect almost all the organ systems. The term ´Inflammatory Pseudo Tumor´ was coined by Umiker and Iverson in 1954, after finding out that the clinical and image findings mimicked that of a malignant lesion. The cause of IPT is unknown. It is characterized histologically by the presence of acute and chronic inflammatory cells with a variable fibrotic response. It is also called spindle cell Pseudotumor, plasma cell granuloma, inflammatory myofibroblastic tumor. The condition is rare in the brain and is usually an intracranial extension of an extracranial tumor arising from the orbit. Rarely, the condition is primarily intracranial or within spinal cord, affecting young men mostly.

Discussion: Inflammatory Pseudotumor is a rare and benign chronic condition of unknown origin. It was first described by Birch-Hirschfield in 1905 for an orbital lesion and was so named by Umiker, et al. in 1954 because of its clinical and radiological similarity to a malignant lesion. Inflammatory Pseudotumor of the head and neck region usually affect the orbit (orbital pseudotumor), orbital apex (orbital apex syndrome), superior orbital fissure (superior orbital fissure syndrome). Extra orbital sites include nasal cavity, nasopharynx, maxillary sinus, sphenoid sinus, infratemporal fossa, choroid plexus, larynx, trachea and skull bone. Extension into the brain parenchyma is rare. IPT of the CNS is rare and are usually seen in children and young adults. Patient usually either has no symptoms or presents with constitutional symptoms such as fever, weight loss and symptoms due to local mass effect from the lesion. In the brain, IPT is either found in the parenchyma, attached to the dura or both. MR Spectroscopy, in such patients, shows a lactate peak which suggests the presence of inflammation and hypoxia and an increase in the choline peak, which suggests increased cellularity and cell membrane synthesis due to inflammation. Histopathology is necessary for a definitive diagnosis of inflammatory pseudo tumor. Biopsy ideally shows nonspecific infiltration of inflammatory cells comprising of lymphocytes, plasma cells, neutrophils and macrophages. Due to the chronic nature of the condition, varying degrees of necrosis is seen. Variable giant cells, calcifications and circular fibrosis of the small veins is also seen. Steroids and surgical resection are the modalities of treatment for inflammatory pseudo tumor of the head and neck. If the tumor is resectable, complete excision is the preferred treatment of choice.

Conclusion: Inflammatory Pseudo tumor is a rare entity. In this too, inflammatory pseudo tumor of the brain is even more rare. There are not many reported cases of Inflammatory Pseudotumor of the brain in literature. It should be considered as one of the differential diagnosis of soft tissue malignant lesions of the brain. Histopathological diagnosis is a must and a confirmative diagnosis cannot be made based only on the clinical and radiological findings. Here we present one such case of inflammatory pseudotumor of the left high parietal cortex.

Biography

Siddharth Gautam has completed his MBBS from the Maharashtra University of Health Sciences, India. He has worked as an Assistant Clinical Researcher in the Department of Neurosciences at Hahnemann University Hospital and Drexel University College of Medicine, Philadelphia. He has completed his MS in General Surgery from Rajiv Gandhi University of Health Sciences, Bangalore, India and MCh Neurosurgery from Chettinad University, Chennai, India. He has authored and co-authored several publications in peer reviewed journals and has participated in many conference presentations. He is currently working as a Junior Consultant in the Department of Neurosurgery at Apollo Specialty Hospitals, Chennai, India.

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