MICRO LABS SYNAPSE

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Neurology Case Series

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Dear Doctor,

Greetings from Micro Labs Limited.

We are happy to bring you "Neurology case series", which contains latest and interesting cases in the field of Neurology.

This issue contains

1. **A Case Report on Tophaceous Vertebral Gout-Related Spinal Cord Compression.**
2. **A case report on Accessory Nerve Ancient Schwannoma.**
3. **A Rare Case of Familial Cerebral Cavernous Malformation.**
4. **A rare case of a sensory trick in a patient with functional dystonia.**

Should you require any specific article or medical information, please feel free to contact us at medicalservices@microlabs.in

Looking forward to your valuable feedback

Best Regards,

Medical Team,

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1. A Case Report on Tophaceous Vertebral Gout-Related Spinal Cord Compression

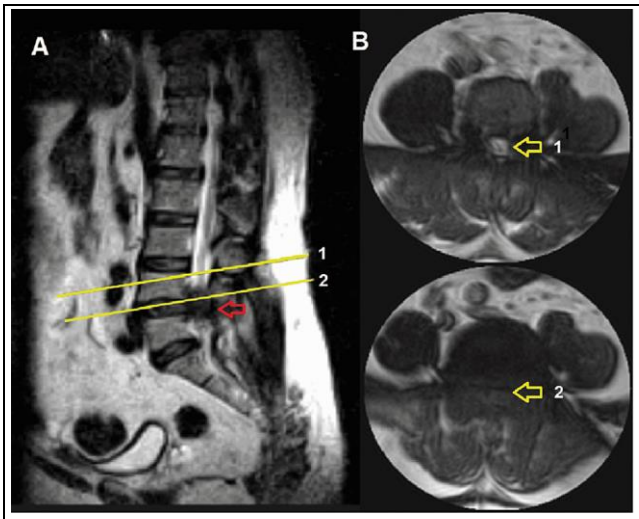
Introduction:

Gout is a frequent and complicated type of arthritis characterised by identifiable signs of inflammation (i.e., dolor [pain], rubor [redness], calor [heat], and tumour [swelling] in the joint). Few cases of spinal gout have been documented in the literature, and spinal involvement is rare.[1] Gout tophi, which typically affect the tiny joints of the feet and hands and cause severe arthritis, are collections of urate crystals in subcutaneous tissues and joints. It is believed that gout arthritis rarely affects the axial skeleton.[2] The clinical, MRI, and pathologic findings of a patient with gout and paraparesis brought on by this atypical gout tophi presentation are described below.

A timely diagnosis and early surgical decompression can decrease or reverse the neurological symptoms in Tophaceous Vertebral Gout patients.

Case presentation:

A 61-year-old man who has had gout arthritis for the previous 15 years visited the neurosurgical outpatient clinic and complained of back pain and losing muscle tone in both lower limbs for several months. In addition, the patient had heart failure, arterial hypertension, and diabetes mellitus. He was aware that he was not adhering to the diet advice or taking the allopurinol, 200 mg daily, as directed by his doctors to treat his hyperuricemia. He thus experienced decreased muscle strength and stiffness in the lower limbs for two months, initially in the left and then the right limb, as well as low back pain with paresthesia for five months. All of these symptoms grew worse over time until they eventually made him unable to walk and carry out daily tasks. Gout tophi were found during the general physical examination in the right knee, right auricular lobe, and elbow and hand joints. His BMI of 34.3 indicated that he was grade I obese. Fasciculations, quick knee and Achilles tendon reflexes, clonus, diminished muscle power (3/5), and spasticity in both lower limbs were all observed during the neurological physical examination. Facet hypertrophy at right T7-T8, left T9-T10, and bilateral T11-T12 levels was visible on axial and sagittal MRI images. The facet hypertrophy protruded into the spinal canal at the level of T11–T12, squeezing the spinal cord. At this level, there were indications of spinal cord ischemia and a constriction of the posterior cerebrospinal fluid column. A hypodense



MRI of the lumbar spine. Sagittal (A) and axial (B) views in T2 sequence. The red arrow shows the tophus as a hypointense mass obliterating the spinal canal. The yellow arrows indicate: 1: axial section above the level of the lesion showing a normal spinal canal. 2: axial section at the level of the lesion showing complete obliteration of the spinal canal.

lesion that filled the spinal canal and narrowed the lateral recesses of the spinal cord was accompanied with facet hypertrophy at the L4-L5 vertebral level. It was believed that these lesions might be connected to underlying gout and account for the neurological symptoms present in the lower limbs based on the patient's history. Consequently, a surgical procedure was suggested. Allopurinol was administered to the patient once they were admitted to the hospital, one dose (100 mg) every eight hours. The procedure was divided into two stages, the first of which involved removing the dorsal lesion at T10–T11 and the second of which involved removing the lumbar lesion at L4–L5. In the initial surgical procedure, a decompressive laminectomy with exeresis of the lesion was carried out. From inside the spinal canal, many brownish-white tumours with a jelly-like texture were removed. The microscopic examination revealed that these lesions were made up of gouty fibrocartilaginous tophi with calcifications and crystals surrounded by inflammatory infiltrates that had invaded the vertebral bone laminae and fibrocartilage. After surgery, the pain subsided but the paraparesis persisted and the muscle strength in both lower limbs remained at 3/5. Thirteen days following surgery, the patient was discharged under pharmacological care that included colchicine (1 mg daily), allopurinol (300 mg daily), and a specific diet. The patient's neurological symptoms did not get any better in the following months. Five months after the first operation, the second one to remove the lumbar lesions was done. Laminectomy procedures were done at the L4 and L5 levels. The medial facet was removed, a foraminotomy was done, and the dural theca was left uninvolved. At this level, a white lesion that appeared to be semi-hard was excised. The bone that had been removed had many greyish fibroelastic fragments with yellowish stippling, according to a macroscopic examination. The microscopic examination revealed fragments of fibrocartilage with crystals and fibromuscular tissue with inflammation, both encircled by an inflammatory infiltrate, which suggested gout tophus. The patient was released with instructions for an outpatient clinic follow-up. Unfortunately, the muscle strength only partially improved despite having intense physical therapy. The patient's hyperuricemia is still strictly under control, and he is still receiving rehabilitative therapy.

Discussion:

The clinical findings and the outcomes of additional tests for a patient with gout tophi in the spinal canal are summarised in the current study. The five clinical forms of gout—asymptomatic, hyperuricemic, gouty arthritis, intercritical gout, and chronic tophaceous gout—are all rather frequent diseases. When hyperuricemic levels are sustained over an extended length of time, as is the case with the chronic tophaceous gout type, gout tophi can form in various joints.[2] Most cases of intraspinal tophi are found by neurosurgeons (19.7%), rheumatologists (19.7%), orthopaedic surgeons (17.6%), and radiologists (17.6%), according to Zhang T et al. Therefore, in order to maximise the early detection of this condition, these specialists should receive the required training. The neurosurgeons suspected intraspinal tophi in the patient who is being discussed here; this suspicion was backed by the radiologists, and it was verified by the pathologists.

Conclusion:

The patient described in this paper had neurological problems that were characteristic of axial gouty arthritis. Although the true prevalence of this condition is unclear, it is definitely larger than is typically thought. Patients with spinal cord involvement related to chronic tophaceous gout must undergo compulsory CT examinations of the spine. The neurological symptoms can be reduced or perhaps eliminated with an early surgical decompression and a prompt diagnosis.

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2. A case report on accessory nerve ancient schwannoma

Introduction:

Schwannomas are benign, well-encapsulated, slow-growing nerve sheath tumours made entirely of Schwann cells that are derived from the neural crest (also known as neuromas, neurinomas "of Verocay," and neurilemmomas). Any myelinated central or peripheral nerve with Schwann cells can give rise to the tumour.[1] Ancient schwannomas are a rare variety of schwannoma that exhibit degenerative alterations as a result of a protracted growth. Researchers have only found one known instance of an ancient variant in the spinal accessory nerve. This is the second instance of this unusual condition; the first affected the XI nerve's intracisternal component.[2]

This is the first report of an uncommon histological variant of schwannoma originating from the XI nerve's intracisternal portion. The rarity of this condition in this could result in an incorrect diagnosis prior to surgery.

Case Presentation:

Due to radiologic findings of two cerebral lesions, a 61-year-old man was sent to our neurosurgery division. His three-month history of gait imbalance, dysmetria, and frequent falls was given to the doctor. He reported three isolated incidents of vomiting but no headaches throughout this time. Hypertension, dyslipidemia, and Type 2 diabetes mellitus diagnoses were part of the individual's medical history. He had also smoked heavily for forty years. The physical examination revealed dysmetria, ataxic gait, bidirectional persistent horizontal gaze-evoked nystagmus, and positive Hoffmann and Babinski signs. The two lesions previously identified were visible on magnetic resonance imaging (MRI). The radiologic diagnosis of suprasellar meningioma was made in the initial mass. The second was a 35 mm by 38 mm by 45 mm extraaxial rim-enhancing cystic lesion. It was situated on the right cerebellopontine angle's inferior portion, extending to the cerebellomedullary cistern, and it caused significant compression on nearby tissues as well as a slight secondary hydrocephalus. Cystic meningioma or an unusual schwannoma were among the differential diagnoses at the time. The patient had cisternal cystic lesion removal surgery with concurrent intraoperative neuromonitoring (IONM). An extended far lateral retrosigmoid craniotomy and a cervical one vertebral right hemilaminectomy were carried out while the patient was seated. Opening the duramater revealed a heterogeneous, highly vascularized cystic lesion that was tightly connected to the right cranial nerves VII, VIII, IX, X, and XII as well as to the brainstem. With the use of IONM, the tumor's origin from the right accessory nerve was confirmed. To protect these structures, a nearly complete excision was carried out. This demonstrated that the mass did not match an ancient auxiliary nerve schwannoma, as was previously believed, but rather a meningioma.

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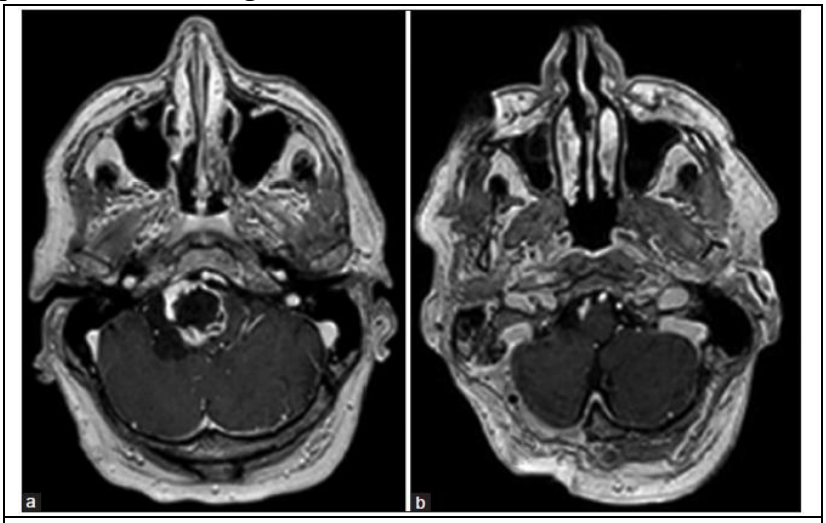
Following surgery, the patient experienced breathing difficulties due to an abundance of respiratory secretions, which led to aspiration pneumonia. His stay in the intensive care unit was extended as a result of this and subsequent cardiac arrhythmias. Another major issue was difficulty swallowing, which necessitated the temporary use of a nasogastric tube for nutrition. Three months following surgery, the patient was discharged having mastered independent oral feeding and deambulation. Initial follow-up procedures were used to treat the tiny capsular fragment that was still present. 13 months following surgery, further MRIs revealed development of the remaining lesion, particularly of its cystic component . The decision was made to reoperate with a debulking objective prior to radiotherapy because of the significant cystic component. The patient experienced a heart arrest of respiratory origin in the postoperative care unit, which led to death after cardiopulmonary resuscitation failed.



Radiological examinations: (a) preoperative axial T1-weighted contrast-enhanced MRI
demonstrating a rim-enhancing lesion. (b) Postoperative axial T1-weighted contrast-enhanced MRI
showing the thin layer of tumor left at surgery.

Discussion:

Schwannomas are tumours that develop from the sheath around a nerve. Meningiomas are the most frequent lesions of the cerebellopontine angle intracranially, and the vestibular nerve is the source of the majority of schwannomas.[2] Most of the time, a CT scan or an MRI is sufficient to reliably diagnose either lesion, but because only 2% of intracranial schwannomas originate from the lower cranial nerves (IX, X, and XI), this can be challenging, particularly when their characteristic morphology is not evident.[3]

Conclusion:

Intracranial ancient schwannomas have been reported in a very small number of patients.To the best of thr researcher's knowledge, this is the first account of a remarkably uncommon histological variant originating from the XI nerve's intracisternal component. The rarity of this condition in this area could result in an incorrect diagnosis prior to surgery.

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3.A Rare Case of Familial Cerebral Cavernous Malformation

Introduction:

Cerebral cavernous malformations (CCMs) are vascular abnormalities of the brain and spinal cord that consist of a single layer of endothelium covering a group of closely spaced, enlarged capillary channels (caverns) that are not yet fully developed and do not have normal intervening brain parenchyma. CCMs have a diameter that can be anything from a few millimetres and many centimetres. Over time, CCMs grow or shrink in size and expand in quantity.[1] It affects up to 0.5% of the population.[2] Here is a case of a man aged 42 years with familial Cerebral Cavernous Malformation

For a precise diagnosis of familial cerebral cavernous malformation, MRI in conjunction with genetic confirmation is crucial.

Case Presentation:

A 42-year-old man came in with a day-long left upper limb weakness and a searing bifrontal headache that had recently started. He gave negative responses to all inquiries about signs of elevated intracranial pressure, had no seizures, and had no systemic symptoms, such as fever or weight loss. He reported no seizures.Patient had a colonoscopy the day before to assess his recurrent diarrhoea, and it revealed several superficial hemorrhagic ulcers in his rectum and large bowel. Regularly, patient ate well-cooked pork. The examining gastroenterologist began metronidazole treatment for a potential case of amoebic colitis, which has a similar endoscopic appearance and clinical course to inflammatory bowel disease. Only the history of patient's other diseases was important: hypertension. his mother's death from a cerebral haemorrhage 15 years' prior made his family history. The blood examination of patient revealed no unusual findings, such as inflammatory markers and eosinophil counts that were within normal ranges and no signs of an infectious condition. Cerebrospinal fluid (CSF) analysis and lumbar puncture were unremarkable. The results of the serology for tuberculosis, amoebiasis, and cysticercosis were all negative. The cerebral hemispheres, cerebellum, and pons all had several nonenhancing, ill-defined hyperdense lesions, and the pons also had prominent focal calcification. Patient was given albendazole for suspected neurocysticercosis after a head CT scan revealed several intracranial hyperdense lesions that resembled calcified neurocysticercosis. The possibility of tuberculoma of the central nervous system (CNS) was also examined, although it was deemed unlikely due to negative results from CSF tests for mycobacterium tuberculosis (MTB), culture, and PCR. His digestive issues persisted despite receiving antiamoebic treatment, and ulcerative colitis was subsequently identified as the cause. Over several years, patient requested more courses of albendazole after expressing subjective improvement. Although brain MRI was not accessible during patient's hospital stay, it was performed serially during his albendazole treatment. Gradient-echo (GRE) MRI revealed widespread numerous minor lesions with blooming anomalies in the cerebellum, brainstem,

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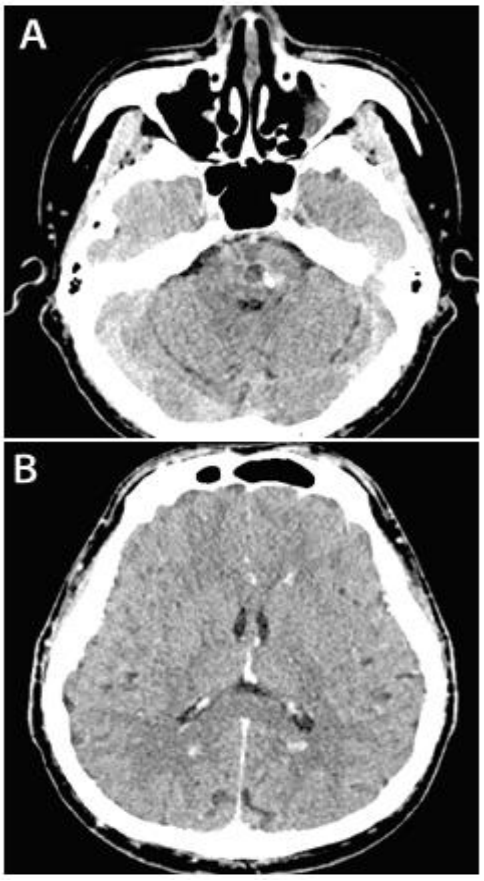
and both cerebral hemispheres . These lesions did not progress over time. There was a clinical suspicion for familial cerebral cavernous malformation due to the absence of radiologic improvement on these and the first negative test for T. solium (FCCM). patient was recommended for genetic testing, which confirmed this diagnosis when it was discovered that patient had the harmful mutation c.334 337del (p.Gln112Phefs*13) in programmed cell death 10 (PDCD10). Over the lateral aspect of the left leg, a few tiny hyperkeratotic, well defined, dark plaque-like lesions were seen, which bled easily when scratched. A biopsy was performed on these lesions, and the results were consistent with angiokeratoma, which is frequently observed in CCM1. Because patient had no significant cerebral haemorrhage, focal neurologic impairments, or seizures, the follow-up care was conservative with frequent examinations.



Brain CT without contrast (A) shows multiple hyperdense lesions in the pons and both cerebral hemispheres that did not enhance on brain CT with contrast (B).

Discussion:

Capillary caverns that are unusually crowded, dilated, and leaky make up cerebral cavernous malformations (CCMs), which are common vascular abnormalities. PDCD10, malcavernin (MGC4607), and Krev interaction trapped 1 (KRIT1), commonly known as CCM1, CCM2, and CCM3, are the three genes that cause FCCM, which is unusual and has an autosomal dominant inheritance pattern. Therefore, it is essential for diagnosis to have a thorough family history, a brain MRI, and genetic testing.

Conclusion:

The common vascular malformation known as a cerebral cavernous malformation (CCM) is made up of improperly grouped, dilated, and leaky capillary caverns.[3] In contrast, FCCM is uncommon and has an autosomal dominant inheritance pattern. PDCD10, malcavernin (MGC4607), and Krev interaction trapped 1 (KRIT1), often referred to as CCM1, CCM2, and CCM3, respectively, are the three discovered causal genes for FCCM. .[1,3] Due to this, correct diagnosis requires a thorough family history, a brain MRI, and genetic testing.

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4. A rare case of a sensory trick in a patient with functional dystonia.

Introduction:

Meige and Feindel, who originally used the term, defined geste antagoniste efficace as movements done by torticollis patients to temporarily stop a dystonic contraction. Even before the corrective manoeuvre was finished, spasm relief could be seen (before the hand touched the face). Thus, they claimed, offered "conclusive evidence of the essentially psychical worth" of these acts.[1] It can be difficult to diagnose and treat primary and functional dystonia separately. For physicians, the presence of a sensory trick can be a valuable indicator, typically indicating a "organic origin" of the condition.[2] The lack of specificity in these manoeuvres is shown by this case study of a sensory trick in a patient with functional dystonia, prompting medical professionals to consider the patient's entire clinical picture.

In dystonia patients, sensory tricks that normally lead in the direction of an organic origin may be diagnostically useful.

Case Presentation:

A 40-year-old man was sent to our neurology outpatient clinic with pain in his head and upper limbs that had been present for three years and was made worse by bending forward and when walking. His medical history included fibromyalgia, anxiety, dyslipidemia, gastro-oesophageal reflux disease, and persistent back pain. The patient said that these motions began suddenly after a work-related incident in which he struck his neck against the bus's steering wheel. Extensive MRI of his head and spine excluded any structural problems. His discomfort and stance worsened with time, forcing him to leave his job as a bus driver. Levodopa had previously been tried by his general practitioner, but it had not been successful.he was not receiving any treatment or medication for his mobility issues. He was never exposed to substances that block or decrease dopamine. The patient displayed an isolated multifocal dystonia during testing, as well as restricted head rotation and modest chin jerks toward his right shoulder. He intermittently displayed improper hand posture with both hands while moving his neck and walking. When the patient gently touched the front of his neck as is usual for geste antagoniste, he noted an improvement in his neck posturing and gait. The examiner inconsistent behaviour.Functional dystonia is only diagnosed based on its clinical manifestations. Primary cervical dystonia, which consists of idiopathic involuntary oscillatory movements with aberrant neck posture, is the main differential diagnosis. was able to

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demonstrate the movements' distractibility as well as the 'sensory trick's' Secondary causes of cervical dystonia, such as cervical sprain, Huntington disease, parkinsonism, multiple sclerosis, and Wilson's disease should be taken into account in order to make the diagnosis of functional dystonia. Pseudodystonia refers to disorders such dystonic tics, atlantoaxial subluxation, congenital muscular, and torticollis that resemble dystonia. Physical examination revealed signs of inconsistency and incongruence. As a result, the dystonia in this patient was functionally neurological in nature and satisfied the criteria for dystonia as defined by the most recent edition of the Diagnostic and Statistical Manual of Mental Disorders (DSM-V). The patient received education, reassurance, and referrals for physical therapy and a neuropsychological evaluation. He accepted the diagnosis and, after working on coping mechanisms, his symptoms began to slightly improve. As a result of his extensive pain syndrome, which includes chronic back pain and contributes to some of his symptoms, he was also sent to the chronic pain services.

Discussion:

The sensory trick is an episodic, voluntarily performed manoeuvre that relaxes dystonic posture and/or movement. Despite the fact that the precise pathophysiological process underlying sensory tricks is yet unknown, their existence is thought to be one of the most significant indicators of an organic cause.[2] The second most prevalent subtype of functional movements, after tremor, is dystonia.[3] The science of medicine and the use of the right language to build rapport with the patient are required while delivering the functional dystonia diagnosis. The role of a neurologist is crucial in validating the symptoms, identifying the illness, delineating its causes, and discussing optimistically biopsychosocial treatment.[4]

Conclusion:

This case illustrates one of the grey areas between functional and organic movement problems, as well as the significance of basing the diagnosis of dystonia on more than just a sensory trick. In dystonia patients, sensory tricks that normally lead in the direction of an organic origin may be diagnostically useful.

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