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Study of Clinical Outcome After Microsurgical Excision of Spinal Haemonagioblastomas

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ABSTRACT
Haemonagioblastomas are benign lesions mostly found in the posterior cranial fossa or the spinal canal. Commonly, the symptoms caused appear to be out of proportion to the size of the lesion due to the associated cysts which can grow significantly in size. As a result, surgical excision of these lesions is quite rewarding in terms of symptom relief. This study undertaken at a high volume tertiary care centre evaluates the demographic characteristics of the patients with such symptomatic lesions, presenting symptoms and post-operative relief of symptoms.

INTRODUCTION

Hemangioblastomas are relatively rare benign tumors of the central nervous system with an incidence of 0.15 per 100,000 population and slight male pre ponderance. These highly vascular tumors are more commonly found in the posterior cranial fossa and spinal cord[1]. These lesions arise from spontaneously or in patients having the autosomal dominant von Hippel Lindau syndrome affecting multiple organ systems[2]. The subset of spinal hemangioblastomas is rare forming somewhere between 2 and 15% intramedullary tumors in various series. Despite their histologically benign character, these tumors cause significant neurological symptoms due to the associated perilesional edema initially and associated cysts later. Microsurgical excision is curative and associated with good outcomes provided en bloc gross total excision is achieved. Surface tumors can be excised with circumferential dissection at the tumor-pia interface but deeper lesions require longitudinal myelotomy. Polar cysts are often found in association with these lesions but aren't neoplastic and do not mandate excision. Lateral tumours can be usually seen and resected by a posterior or posterolateral approach whereas anterior lesions in between the two ventral root entry zones require an anterior approach with a corpectomy. Here, we present our experience of microsurgical excision of 8 cases of spinal Haemonagioblastomas including post-operative neurological improvement.

MATERIALS AND METHODS

Summary of Patients: Eight patients underwent microsurgical excision of spinal Hemangioblastomas between June 2020 and June 2024 by the senior author. 5 men and 3 women with aged between 24-43 years were operated in the period of the study after contrast MRI scanning of the neuraxis as well as adequate screening of the patients for other lesions outside the neuraxis.

Surgical Technique: All the lesions were approached posteriorly after putting the patient in a prone position. After laminectomy extending beyond the rostro-caudal extent of the lesion and durotomy, the lesions were separated through a natural plane from the neural tissue with minimal initial disturbance to the vascularity to maintain the natural colour contrast. En masse excision was achieved by such technique taking care to subject the neural tissue to minimum amount of traction possible. Watertight dural closure with tissue glue reinforcement ensured non-occurrence of post-operative CSF leak in any of the cases[3-7].

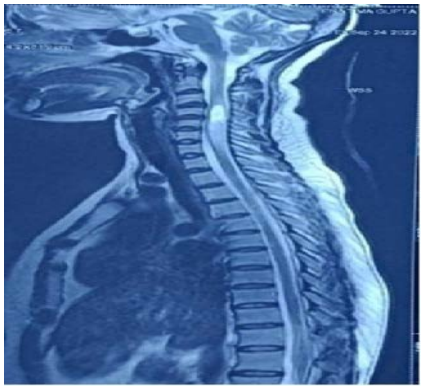

Fig. 1: Watertight Dural Closure with Tissue Glue Reinforcement

Post-operatively the patients were followed up on out-patient basis after discharge to observe neurological improvement, complications or recurrent symptoms and signs. Follow-up was done monthly for the initial 3 months and then 6 monthly thereafter. The patients' functional neurological status per-operatively and post-operatively was classified by Nurick grading for comparison.

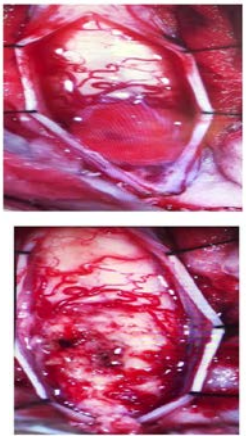

Fig. 2: Neurisk Grading

All the tumours were found to be intramedullary. 3 of these patients were found to have von Hippel-Lindau syndrome with multiple lesions in the neuraxis. The duration of pre-operative symptoms ranged between 3 months-8 months. Sensory disturbance was the most common initial symptom in the form of numbness and parenthesis which was neurologically consistent with the level of the lesion. 5 patients had prominent accompanying pain.

RESULTS AND DISCUSSIONS

Patients were found to be between 24-43 years old with 6 of them in the third decade with the average age as 30.25 years. Among the 8 patients, 5 were male and 3 female. 3 patients were diagnosed with von

Sr. No.	Age (Y)/ Sex	Tumour Level	Symptoms	Clinical Grade		Complications	Follow-up duration (months)
				Pre-operative	Post-operative		
1	24/M	C7-D1	Numbness, Weakness	IV	II	None	48
2	37/F	C6	Numbness, Backache	II	II	None	42
3	29/M	D5	Numbness, Backache	II	II	None	36
4	27/F	C6-C7	Weakness	IV	II	None	30
5	43/M	D2-D3	Numbness, Backache	II	III	Neuropathic Pain	24
6	28/M	C7	Numbness, Weakness, Backache	III	III	None	24
7	25/M	D4-D5	Numbness, Backache	II	II	None	18
8	29/F	C6-C7	Numbness, Weakness	IV	II	None	12

0	Signs or symptoms of root involvement but without evidence of spinal cord disease
1	Signs of spinal cord disease but no difficulty in walking
2	Slight difficulty in walking which did not prevent full-time employment
3	Difficulty in walking which prevented full-time employment or the ability to do all housework but which Was not so severe as to require someone else's help to walk
4	Able to walk only with someone else's help or with the aid of a frame
5	Chair bound or bed ridden

Hippel-Lindau syndrome on account of multiplicity of neuraxial lesions. None of these patients showed extra-neural lesions on screening consistent with the syndrome. The most common symptom was sensory deficit or numbness. All the patients underwent microsurgical excision and gross total excision was achieved. Only one patient reported neuropathic pain post-operatively which was nearly completely relieved by drugs. All the patients reported slight symptomatic improvement in terms of decrease in numbness and backache immediately in the post-operative period on being questioned. The motor deficits showed significant improvement on more prolonged follow-up within about 6 months.

Epidemiology and Clinical Features: A male perponderence has been reported for cases of spinal Hemangioblastomas with the ratio varying between 1.6:1 and 5.5:1. In our series it appears to be 1.67:1. These lesions were more commonly observed to occur in the cervical and dorsal region but more rarely in the lumbo-sacral dorsal roots or filum terminale. Here, 50% lesions occurred in the cervical region, 1 from the cervico-dorsal junction and 37.5% in the dorsal region. The associations of spinal haemangioblastoma lesions with von Hippel-Lindau syndrome has been reported to be quite strong: between 20-45% which turned out to be 37.5% in this series. vHL syndrome is an autosomal dominant neoplastic disease resulting from the germline mutation of VHL tumour suppressor gene that causes the development of several lesions throughout the body. The treatment of spinal haemangioblastomas can be complicated by the presence of other lesions in different organs as pheochromocytomas. vHL syndrome patients also tend to present with Hemangioblastomas lesions at an earlier age than sporadic cases. Management also differs in syndromic and sporadic cases. Sporadic cases are usually symptomatic at presentation and surgical excision is the primary option. Multiple neuraxial lesions are considered to be symptoms of a wider disease that does not have a complete cure at this time. Hence, excision of only frankly symptomatic lesions or those showing significant pathogenesis progression is advocated in syndromic cases. Around 80-90% Haemonagioblastomas have been reported to be associated with cysts which can cause misleading clinical features localizing to distant spinal levels than the actual lesion itself. Symptoms have been reported to vary in severity from mild sensory or motor deficits, pain, dysesthesia, incontinence or even bulbar symptoms from high cervical lesions. Sensory dysfunction or pain has usually been reported as the initial symptom. Use of clinical grading scales has often been found to be useful to document and report long term improvement on follow-up.

Radiographic Characteristics: Contrast MRI was done for all the patients showing the characteristic appearance of a small T1 isointense T2 hyper intense nodule showing homogenous contrast enhancement. Angiography and endo vascular embolization often aids in safe resection of these lesions on account of high vascularity but wasn't possible in our setup.

Characteristics of Lesion with Surgical Relevance: The anatomical location of spinal Haemonagioblastomas has profound relevance for the operative technique and approach required for successful excision. Around 90%-100% lesions have been founud to be associated with the dorsal side of the cord with around two-third lesions being associated with the dorsal root entry zone. These findings from previous cases were confirmed in our study with all the lesions being situated dorsally.

Clinical Outcome After Microsurgical Excision: 3 patients (37.5%) with more neurological deficits and

poorer pre-operative Nurick grading showed significant post-operative improvement with improvement in functional grading by 2 grades. 4 patients (50%) showed no significant change in post-operative period with gradual improvement over prolonged follow-up. 1 patient (12.5%) showed functional deterioration in spite of clinically preserved motor function on account of severe neuropathic pain which improved gradually over time with the subsidence of the pain with drugs.

CONCLUSION

Spinal Haemonagioblastomas are surgically curable lesions. Gross total excision achieved with meticulous microsurgical technique and controlled intra-operative blood loss leads to minimal neurological morbidity and a good outcome.

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