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Low back pain is a frequent complaint in PM&R OPD. Tumors of the cauda equina play an uncommon but nevertheless important role in the "sciatic syndrome". Early detection will lower the risk of neurological sequelae. However, on clinical grounds alone, it is sometimes difficult to differentiate between a cauda equina tumor and a prolapsed intervertebral disc. Recent advances in imaging techniques had increased the diagnostic rates. This study was based on 6 cases of ependymoma of the cauda equina, collected from January 1986 to May 1992 at VGHTC. Three of them were detected at PM&R OPD. The clinical symptoms, physical examination, treatment and outcome will be discussed.

The initial symptom is back or buttock pain (100%), while one case was combined with headache, vomiting and neck stiffness which proved to be subarachnoid hemorrhage later. The late symptoms include sensory deficit (100%), motor deficit (50%) and sphincter disturbance (67%). Nocturnal pain (67%), which is rare in herniated intervertebral discs, is a traditional feature of the cauda equina tumor.

MRI or CT-myelogram are efficient diagnostic tools.

After surgical excision with or without radiotherapy, most symptoms relieved. Two out of four cases with preexisting urination problems improved after surgery. Two cases developed difficulty in urination post-operatively. After bladder training, they regained their function within 4 weeks.

Key words: ependymoma, cauda equina, back pain

INTRODUCTION

Tumors of the cauda equina are an uncommon but nevertheless important cause of the "sciatic syndrome" which is frequently encountered at PM&R OPD. At times, it is quite impossible to differentiate, on clinical grounds alone, between a cauda equina tumor and a prolapsed intervertebral disc. However, recent advances in diagnostic and imaging techniques have vastly expanded our capability to detect lesions early. To take full advantage of this new ability, the clinician must be equipped with a sufficient index of suspicion and adequate knowledge of the natural history of the disease in the differential diagnosis.

With this in mind, we reviewed our experience with ependymoma of the cauda equina at VGHTC. Six patients had been treated since 1986, and three of them were detected at PM&R OPD. The diagnosis, treatment, and outcome of these patients are retro-

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spectively reviewed with the goal of identifying potential for improved therapy for this disease entity.

PATIENTS

We collected patients from January 1986 to May 1992 with the diagnosis of ependymoma of the cauda equina region. Total number is six patients, including 3 males and 3 females; their average age at presentation was 26.5 years (range 14 to 48 years). Follow-up periods are from 1 month to 6 years and 4 months (Table 1).

4 patients had first been seen in Orthopaedic or Rheumatology clinics where nonspecific low back pain was told. 3 patients were eventually diagnosed as tumor in our OPD, 2 in Neurology department and 1 in Neurosurgery OPD.

RESULTS

Of all these 6 patients, all presented initially as back or buttock pain while 1 (No.2) combined with headache and vomiting which proved later to be subarachnoid hemorrhage due to tumor bleeding.

Pain may be intermittently sustained and slowly worsening, or continuously progressing from onset. At first, the pain was localized at the back or buttock. Sciatic pain, especially bilateral, developed later. Sensory deficits were complained of in 6 patients (100%), while motor deficits in 3 patients (50%), ranging from mild leg weakness to foot drop. In 4 patients, the pain worsened at night by lying down and was relieved by walking around or rolling over the bed. 2 patients received physical therapy and 3 took analgesics without relief. Sphincter disturbance was noted in 4 patients which maybe an indication of late involvement.

Limited motions of the lumbar spine were shown in 4 patients, especially flexion (Table3). Initially, motor and sensory disturbances were infrequently present. However, at the time of diagnosis, neurological dysfunctions were seen in all patients. Motor deficits were seen in 5 patients, while 4 showed bilaterally. Sensory deficits were in all 6 patients with multiple dermatome involvement in 5 patients. Deep tendon reflex, like ankle or knee jerk, diminished in 4 patients, but increased in 1 patient. Diminution of straight leg raising was noted in 5 patients, bilaterally in 4 cases. Lax anal tone was found in 3 patients with 1 (No.1) showed absent bulbocavernous reflex.

The definite diagnosis was made by CT-myelogram in 3 patients, another 3 cases by MRI. Lumbar punctures in the patient presenting as subarachnoid hemorrhage revealed high protein content (144-375 mg/100ml).

All patients underwent surgery performed by different qualified neurosurgeons. The operation consisted of one or more laminectomies followed by intraspinal exploration. The tumors, all situated intradurally, could be completely removed in 5 patients. Too much adherence of the tumor of the cauda equina roots was noted in the remaining 1 patient. Only subtotal removal of the tumor was done, which was followed by radiation therapy (tumor dose 5250 rads).

OUTCOME

After operation, pain improved in all patients. Motor deficit deteriorated in 1 patient. Sensory disturbance improved in 4 patients, stationary in 2 patients. As to micturation problem, 2 remained the same as pre-operative condition; while 2 developed post- operatively, but resolved within 4 weeks after bladder training (Table 1). Urodynamic study in the above mentioned 4 patients showed detrusor areflexia or acontractile urinary bladder (Table 4).

Table 1. Clinical date of 6 patients treated for an ependymoma of the cauda equina

Case No.	Sex	age at time of treatment	duration of complaint	removal	R/T	follow up	results of treatment			
							pain	motor deficit	sensory deficit	micturation
1	M	14	8M	total	−	6Y4M	1	1	2	2
2	F	16	1Y	total	−	2Y8M	1	0	1	1
3	F	22	9M	total	−	1Y10M	1	1	1	1
4	M	35	10Y	partial	+	10M	1	3	2	2
5	F	24	1Y1M	total	−	1M	1	1	1	4
6	M	48	4Y7M	total	−	1M	1	1	1	4

Legend: F = female M = male
R/T=radiotherapy
0 = nor present before, neither after operation
1 = improved
2 = no further deterioration
3 = worsened
4 = only present after operation

Table 2. Symptoms at time of onset and examination

symptom	no. of patients	
	onset	examination
back pain	4	4
buttock pain	2	2
sciatica		4 (bilateral in 4)
sensory deficit		6
motor deficit		3
night pain		4
sphincter disturbance		4

Table 3. Signs at time of examination

sign	no. of patients
limited motion of L-spine	4
motor deficit	5 (bilateral in 4)
sensory deficit	6 (multiple dermatomes in 5)
diminished DTR	4 (increased in another 1)
postivie SLRT	5 (bilateral in 4)
lax anal tone	3

Legend:
DTR = deep tendon reflex
SLRT= straight leg raising test

Table 4. Post-operative urodynamic study and residual urine check-up

case no.	urodynamic study	residual urine (ml)	remark
1	acontractile UB	10	Crede maneuver
4	detrusor areflexia	100	Crede maneuver + Duvoid(25) 1# BID
5	detrusor areflexia	35	Duvoid (50) 1# TID → DC
6	detrusor areflexia	30	Duvoid (50) 1# TID → DC

DISCUSSION

Ependymomas are uncommon intracranial lesions in the adult group, comprising approximately 5% to 6% of all brain tumor [1,2]. In the spinal cord, however, they represent approximately 63% of all parenchymal tumors, although figures have ranged from 30% [3-5] to 88% [1,6,7]. Ependymomas of the cauda equina region represent one-half or more of all intraspinal ependymomas [4], and to present most after in the third decade, which is older than our series (average 26.5 years).

Kernohan et al. [8] described three histological types and later proposed further classification of ependymoma into 4 grades [9], similar to the grading system for astrocytoma. However, despite some variation in cytologic features and the presence of atypia and modest mitotic activity in most cases, the gross

characteristics of the tumors appear to be of greater prognostic significance than the histological features [10].

Ependymoma outside the central nervous system is rare [11]. Helwig and Stern [12] divided sites into four general situations: (i) metastases or direct extension following surgical excision from the central nervous system primary tumor; (ii) direct extension to the soft tissue of the sacrococcygeal area from a primary ependymoma of the lower spinal cord, cauda equina, or filum terminale; (iii) occurence as a primary presacral, pelvic, or abdominal tumor; and (iv) as a primary tumor of the skin and subcutaneous tissue of the sacrococcygeal area without any demonstrable connection with the spinal cord.

Although likely to recur locally, ependymomas primary in the cauda equina region rarely metastasize systemically [13]. In contrast, those far less common examples arising in the extraspinal locations more frequently do so [13]. However, intracranial tumors resulted from retrograde seeding via the CSF had been reported [15]. In our series, there were neither local recurrence nor metastasis during the follow-up periods.

The tumors usually present as a slow-growing intradural mass. In our study, the duration of complaint ranges from 8 months to 10 years. Reviewing the classical signs of the cauda equina lesions, Wechsler [15] noted that the cardinal symptom is pain, which may continue for a long time before neurological signs appear, so that the tentative diagnosis of hysteria is sometimes made. The pain is mostly intermittent in the early phase [16] and progressive in a later stage, as was seen in all of our patients. Impassiveness of pain to conservative treatment is another differential characteristic [18], as 2 patients in this study. 4 patients developed bilateral sciatic pain at the time of diagnosis, which is an absolute indication for myelogram [17]. If it is of sudden onset, subarachnoid hemorrhage from degeneration of a tumor is suspected (Falconer 1966).

A spinal etiology of subarachnoid hemorrhage seems to be very rare (about 0.6 to 1.0% according to Perel et al. [18]). One patient in our study had acute onset of severe headache with vomiting and menigitis was first impressed by the Infection department. she also described a mild degree of buttock pain of 1/2 month duration. Lumbar punctures showed xanthochromic CSF and elevated protein content (144-375 mg/100ml). Physical examination revealed menigeal signs and local tenderness over the sacral area. Neck stiffness improved with conservative treatment but buttock pain persisted. Finally, an intraspinal tumor at the cauda equina was found by MRI. She received surgical excision with improvement.

Nocturnal pain is a traditional feature of the cauda equina tumor. Dodge et al. (1951), Craig et al. and Fearnside and Adams (1978) repored an incidence of approximately 50%. In our series, 4 patients (67%) had night pain. Sphincter disturbance is mostly considered as a late complication, although early occurence has been noticed [17]. 4 of our patients complained of urination problem before the operation. 2 improved post-operatively while 2 remained the same. Interestingly, 2 patients (No.5,6) developed difficulty in urination following the surgery. Urodynamic study showed detrusor areflexia. After bladder training, the symptom subsided within 4 weeks. Operative hemorrhage and edema may be the cause.

The lax anal tone was noted in 3 patients, thus reemphasising the importance of rectal examination in the assessment of patients with back pain. The daily activities were restricted in 2 male patients due to flaccid bladder. Crede maneuver is the only way to empty the bladder. Early recognition of the tumor cannot be overemphasised to prevent the neurological sequelae.

MRI is the initial study of choice [19], but when it is negative in the face of persisting symptoms, myelogram with CT must be performed [20], which is also a useful tool in the follow-up study. In our series, there is equal diagnostic rate between MRI and CT-myelogram.

All patients having ependymomas of the cauda equina region should undergo the maximum surgical resection compatible with preservation of the preoperative neurological status. With total resection of the tumor, the survival rate could be 85% to 100% [11]. Subtotal resection results in recurrence or metastasis [21]. Radiation therapy is recommended for lesions not totally resected or recurrence [21]. In our study, 5 patients received gross total removal of tumor without additional radiation therapy. The one with partial removal of tumor received radiotherapy (5250 TD). They all lived well till now.

In summary, ependymomas of the cauda equina region are rare tumors, which have a generally good prognosis in contrast to similar tumors araising ectopically outside the central nervous system or to intracranial ependymomas. Early detection enhances the chance of reversibility of the neurological disturbance and the possibility of complete excision. The course and nature of the pain will be helpful to the differential diagnosis with lumbar disc syndromes. It is recommended that all patients suffering from a back pain or sciatic syndrome which reacts insufficiently to conservative treatment should undergo further study, like MRI of CT-myelogram. The follow-up study is also important for the possibility of local recurrence of metastasis.

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"下背痛"在復健科門診是頗爲常見的主訴，腫瘤雖佔其中一少部份，但卻是最爲重要的一部份。倘能早期發現，必能降低神經方面的後遺症，而在臨床症狀上，有時往往不易區分椎間盤凸出與馬尾腫瘤。但隨著診斷性工具的進步，早期診斷率已大爲提高，本篇報告台中榮民總醫院復健科自1986年1月至1992年5月收集的6例馬尾區室管膜瘤病人，其中3例在復健科門診診斷出。以其臨床症狀，理學檢查、結果及後遺症加以分析討論。

初期臨床症狀以背痛或臀部痛居多（100％）。但有一例合併頭痛、嘔吐、脖子僵硬，經檢查診斷爲室管膜瘤合併蜘蛛膜下腔出血。後期則出現各種神經症狀，如感覺失常（100％），下肢無力（50％），及括約肌的問題（67％）。夜間疼痛（67％）是馬尾腫瘤的特徵，一般椎間盤凸出少有之。

核磁共振造影或電腦斷層合併脊髓造影爲有效的診斷工具。經手術治療或合併放射治療，大部份的症狀皆獲緩解。四例開刀前有小便困難，開刀後2例有進步。兩例在開刀後才發生此問題，經膀胱訓練於四週內復原。

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