spine disclosed two intradural extra-
medullary masses located anterior to the
spinal cord at the C7-T1 level displacing the
cord posteriorly. These two masses had a higher signal intensity compared with
CSF on both T1 and T2 weighted sequences (fig 1).

The patient was subjected to C6-T1 laminectomy. Before opening the
intraoperative sonography showed two sep-
rate cystic lesions. After severing the den-
tate ligaments, the spinal cord was retracted, exposing the cysts. The one
behind the C7 vertebra was attached to the
pia of the cord, was ovoid in shape, and
partially invaginated into the cord. The sec-
ond cyst, behind the T1 vertebra, was attached to the dura anteriorly and was
pul-
satile. It caused no obvious cord compres-
sion. These two cysts were extracted and excised from their attachments using an
operating microscope.

The patient made an uneventful recov-
ery. The paraesthesia over his chest and
right leg disappeared shortly after the opera-
tion, whereas weakness and atrophy of both
his hands showed gradual but steady in-
creasing during follow up to six months
after the operation.

Each cyst measured 1.5 cm in diameter, had a light blue tint, and contained milky
fluid. Histologically, both cyst walls were
composed of connective tissue lined by a
single layer of ciliated or non-ciliated
columnar epithelium. Intracytoplasmic
mucin was identified in some of the epithe-
lium lining with alcin blue and mucicarmine, and was positive to a diastase
pretreated periodic acid Schiff (DPAS)
reaction. Carcinoembryonic antigen was
demonstrated immunohistochemically in
cells (fig 2). As the enteric tract is posi-
tive for this strain from the second to the
sixth month of fetal development, Miyagi et
al have reported that the epithelial cells of
intrapial enterogenous cysts respond posi-
tively to staining for carcinoembryonic
antigen.

Intraspinal enterogenous cysts usually
affect young men more than women. They
have been found more often in the cervical
than in the thoracic or lumbar region. They
usually occupy the intradural extra-
medullary space. The initial clinical mani-
festations of our patient were weakness and
atrophy of the intrinsic muscles of his right
hand for 15 years without any sensory
involvement, clinical features rarely seen in
the previously documented cases. Woo and
Sharr have subgrouped the cysts into two
types—the developmental type usually
located anterior to the cord in the cervical
region and the teratomatous type usually
located posterior to the cord around the
conus.

Intraoperative sonography showed not
only the extent of cord compression, but
also the coexistence of a second cyst, which
might not have been easily seen during the
operation especially when the cyst is ante-
riorly located and invaginated into the
cord.

Table 1: Craniopharyngioma: survival of more than 10 years

<table>
<thead>
<tr>
<th>Case No</th>
<th>Age (y)</th>
<th>Sex</th>
<th>Survival (y)</th>
<th>Status</th>
<th>Remarks</th>
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<tr>
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*Myocardial infarction; †encephalitis; ††hepatoma; Be = bleomycin; Rad = radiation; Op = operation.

Table 2: Craniopharyngioma: survival of between five and 10 years

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*Brain tumour; †heart disease; ††rectal cancer; Be = bleomycin; Rad = radiation; Op = operation.
and 10 years (group B) and those surviving for more than 10 years (group A). The longest period of follow up in Group A was 39 years and the minimum follow up period was 16.7 years. In group A there were three deaths, of which two were due to unrelated causes. One year before the death of case 3, he had undergone a high fever and a CSF leak was obtained by lumbar puncture. Encephalitis Japonica was diagnosed, he was given no specific treatment, and was discharged from hospital with unsteadiness of gait and subsequent convulsive seizures. One year later he complained of sudden headaches and convulsive seizures. He lost consciousness and died and there was no necropsy.

In group B (14 cases who survived between five and 10 years) 10 had died, eight due to recurrent tumour and two due to unrelated causes. The average age at operation and male/female ratio was 27.2 years and 8/7 in group A and 27.1 years and 10/4 in group B. The same surgical technique were used in both groups with 10 of 15 cystic tumours in group A and nine of 14 in group B. Histologically, five cases were adamantinomatous, seven were squamous, and three were not determined in group A, and in group B six were adamantinomatous, seven were squamous, and two undetermined.

The NOR number of group B was determined in 19 cases from their paraffin embedded specimens. All cases had a low proliferative potential (NOR 1-76 (SD 0-21)) with no differences between the groups. (group A 1-73 (0-13), 10 cases; group B 1-80 (0-27), nine cases).

It is well known that optic or hypothalamic glioma, cerebellar astrocytomas, ganglioglioma, and some non-glial benign tumours display spontaneous cessation of their growth after an active proliferative growth period but such a phenomenon has not been documented for craniopharyngioma. Among the 15 cases who survived more than 10 years in our series, there was no death due to tumour recurrence or regrowth. By contrast, among those 14 cases with less than 10 years survival, 10 patients died due to recurrence of the tumour. There are many statistical survival studies for craniopharyngioma but few dealing with long term survival.8,9 Each of these shows that there is a risk of recurrence once patients survive for 10 years. For example, in the series of Yasargil et al., patients surviving 11 to 22 years after subtotal surgery, no death occurred in 30 children and 33 adults—90% of these cases underwent total removal. Jose et al reported the follow up results of limited surgery and radiation therapy in 173 cases of craniopharyngioma with a median follow up of 12 years. The 10 and 20 year progression free survival rates were 83% and 79% and actuarial 10 and 20 year survival rates were 77% and 60%.

In conclusion, our study supports other evidence in the literature that there may be spontaneous regression in the active proliferative potential of some craniopharyngiomas that is not predictable by the Ag-NOR technique. Of course, some craniopharyngiomas may always have a low proliferative potential and it is patients with these who survive for more than 10 years. Our data lend support to the policy of subtotal removal and subsequent adjuvant therapy in selected patients but a method for defining this sub-group remains unresolved.

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Although these findings clearly show that chronicity has an adverse effect on adjustment to epilepsy overall, the following areas were not affected either by employment, attitudes towards the medical profession, attitudes towards taking medication, and difficulties with the family. In these areas, seemingly unaffected by the experience of having epilepsy over a period of time or by the experience of a larger number of seizures, other dynamics must be involved in causing problems with adjustment, for instance the effect of the stigma of diagnosis. In different patients, however, factors vary widely and responses to the condition vary in a complex manner. Interventions, therefore, designed to improve psychosocial adjustment, require a high degree of complexity and flexibility.

The findings from this study are important for two reasons: firstly, they illustrate that a potential factor affecting psychosocial adjustment is chronicity; secondly, they emphasise that in most areas stigma is not inherent in the condition, and furthermore that there are wide individual variations. We have focused on chronicity but there are of course other factors involved in the process of adjustment. The problems of psychosocial adjustment are clearly relevant

The table shows the percentage of patients who indicated a moderate or severe problem of adjustment in each of the 14 areas comprising the psychosocial adjustment profile. Variable means were compared with t-tests. A significance level of p < 0.05 was taken to indicate an important difference between groups.

Clearly, epilepsy, or at least having seizures, can be very distressing for the patient; the inventory not surprisingly showed that most patients with epilepsy have problems of psychosocial adjustment. Only in one area, however, did significant difficulties occur in more than 50% of the newly diagnosed group, this was the fear of seizures; in the chronic group, difficulties occurred in more than 50% of patients in nine of the 14 areas. The chronic group were more worried about their future, had a more negative outlook on life, had problems in relation to social life and leisure activities, and were more likely to feel socially isolated.
Craniopharyngioma: early and long term recurrence after partial removal.

A Nishimoto, T Matsuhisa, K Kunishio, T Maeshiro, T Furuta and T Ohmoto

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