Corticosteroid therapy in encapsulating peritoneal sclerosis

Sir,

Among the complications related to PD, the most serious and life-threatening one is encapsulating peritoneal sclerosis (EPS), with a reported mortality rate of more than 60% within 4 months of diagnosis [1–3]. The poor prognosis of EPS has had a negative impact on the acceptance of PD as an adequate alternative dialysis therapy to HD. This trend has been especially prominent in Japan where there are a substantial number of long-term CAPD patients. Nonetheless, a few recent reports have suggested that immunosuppression and/or corticosteroid therapy might be effective in the treatment of EPS [4,5]. Acting on these reports, the Japanese Ministry of Welfare decided in 1995 to establish a working group on the definition, diagnosis, and treatment of EPS [6]. The working group concluded that corticosteroids were potentially effective in the treatment of this serious complication. Encouraged and motivated by the above-mentioned recommendation, we decided to launch corticosteroids as a first line therapy for EPS in early 1997.

Table 1 summarizes the clinical outcome of EPS in all patients who developed EPS between the period before and after the year of 1997. The diagnosis of EPS was made based upon the diagnostic criteria proposed by the working group [6]. Briefly, the definition of EPS is a clinical syndrome associated with ileus symptoms such as nausea, vomiting, and abdominal pain due to a mechanical bowel obstruction derived from irreversible thickening and/or sclerotic changes of the peritoneal membrane. Of the 205 patients treated for CAPD over a period of 18 years (1981–2000), 11 patients (5.5%), six before late 1996, and five after early 1997, were diagnosed as having EPS. The duration of CAPD at the time of diagnosis for patients overall was 51 ± 27 months. All patients had an episode of bacterial or fungal peritonitis at least once while they were maintained on CAPD. Before 1997, the basic therapeutic strategy for EPS had been to sustain the rest of the bowel and to apply total parental hyperalimentation (TPN). After early 1997, corticosteroid (prednisolone 0.5 mg/kg/day), was given orally to the patients with EPS in addition to the conventional treatment. Before late 1996, all the patients not subjected to corticosteroid died of EPS related complications within 8 months

Table 1. Clinical outcome of the treatment for ESP before and after the introduction of corticosteroid therapy

Patient	Age (years)	Sex (m/f)	Duration of CAPD (months)	Peritonitis	UF loss (D/PCr)	PSL (mg) (initial)	PSL (mg) (now)	Prognosis
Before 1997								
1 H.N.	38	M	49	Pseudo, Candida	Yes (not done)	No	No	Deceased
2 T.A.	64	F	10	Pseudo, Candida	Yes (0.86)	No	No	Deceased
3 Y.Y.	61	M	50	Unknown	Yes (0.96)	2No	No	Deceased
4 T.K.	58	M	15	Pseudo	Yes (0.89)	No	No	Deceased
5 M.S.	61	M	62	Pseudo	Yes (not done)	No	No	Deceased
6 T.L.	41	F	104	Pseudo	Yes (0.91)	No	No	Deceased
After 1997								
7 K.S.	57	M	57	MSSA	Yes (0.84)	40	off	Alive and well
8 M.H.	36	F	45	MRSA	Yes (0.69)	40	5	Alive and well
9 H.K.	29	M	56	MRSA	Yes (0.90)	40	5	Alive and well
10 T.T.	57	M	77	Unknown	Yes (0.86)	30	5	Alive and well
11 A.T.	38	M	30	Unknown	Yes (0.94)	30	10	Alive and well

Pseudo: *Pseudomonas aeruginosa*; Candida: *Candida albicans*; PSL: Predonsisolone.

of diagnosis. In contrast, after 1997, all the patients treated with corticosteroid are all alive and well today, maintaining good prognosis for at least 1–3 years after the diagnosis of EPS. Only case 7 underwent surgical removal of the thickened omentum while undergoing corticosteroid therapy.

Junor *et al*., in 1993, noticed from their clinical observation of 17 patients with EPS that patients receiving immunosuppressive drugs or recipients of kidney transplantation showed good results [5]. Similarly, Hawley *et al*. indicated that one EPS patient showed a dramatic improvement after kidney transplantation [7]. Mori *et al*. reported that corticosteroid alone was dramatically effective in patients with EPS [4]. In addition to the conventional treatment for EPS including cessation of PD, oral fasting, and TPN, surgical treatment might be taken into consideration as one of the choices [8,9]. However, there has been no agreement as to the surgical management of EPS at present. Generally, surgical treatment to remove the adhesive lesion is extremely difficult, as well as being very hazardous. Surgical management should, therefore, be applied only as deemed necessary. Of note, however, is the suggestion by Bhandari *et al*. that immunosuppression in EPS improves operability and therefore should be considered as an initial treatment with a view to subsequent surgery [10].

The pharmacological mode of corticosteroid action on EPS is, at present, largely unknown. However, one could speculate that it may be via both the anti-inflammatory effect and the immunosuppressive effect of this agent. One must be aware that corticosteroids must be used for PD patients developing EPS under consideration that the patients' treatment would not be complicated by an active infection.

Noticeably, our present study shows that EPS does occur even in a CAPD patient with only 10 months duration in which *Pseudomonas aeruginosa* and Candida were identified as causative microorganisms (Table 1). The evidence clearly refutes the notion that long-term use of peritoneum is associated with peritoneum progressing into fibrotic/sclerotic/adhesive changes which would lead to EPS. Pseudomonas alone or a mixed infection with Pseudomonas and Candida may be strongly associated with an abrupt development of EPS. This also raises the need to be fully aware of the possible development of EPS, even if the duration in which patients are maintained on PD is short.

In summary, our data suggest that therapy with corticosteroids is effective in PD patients with EPS, and that steroid therapy should be considered as a first line therapy. Early recognition of EPS by constant clinical, radiological and ultrasonic surveillance, and prompt withdrawal from PD also appears to be of importance in contributing to better results. EPS seems to be no longer a fatal complication if timely cessation of PD and appropriate use of steroids can be achieved.

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