Comments

The study shows that high doses of 5-amino salicylic acid and sulphasalazine were equally effective in maintaining remission in patients with ulcerative colitis, but the increased dose appeared to have no real advantage over the lower dose of 1.2 g previously used. Higher serum concentrations of 5-amino salicylic acid and acetyl 5-amino salicylic acid were recorded in the serum of patients taking the oral 5-amino salicylic acid preparation compared with those taking sulphasalazine. It is possible that the higher doses might be of value in treating acute colitis.

We thank Boots Pure Drug Company, Nottingham for supplying placebo sulphasalazine, and Rohm Pharma GMB for preparing the tablets of 5-amino salicylic acid. Inquiries about 5-amino salicylic acid should be sent to Tillotts Laboratory, Henlow, Bedfordshire.

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Screening for neural tube defect: false positive findings on ultrasound and in amniotic fluid

Combining detailed ultrasound scanning with examination of amniotic fluid increases specificity in the diagnosis of neural tube defect. We report a case with a very high amniotic fluid α fetoprotein concentration, a positive acetylcholinesterase band, and an ultrasonic pattern strongly suggestive of encephalocele but which on termination was found to be a case of Turner's syndrome.

Case report

A 23 year old woman (gravida 1) underwent amniocentesis at 17 weeks of gestation because of anxiety over a borderline serum α fetoprotein concentration (2.3 multiples of the median) and because she was receiving naproxen for a rheumatic condition. A clear straw coloured specimen of amniotic fluid was obtained and the α fetoprotein concentration estimated as 2000 mg/l (reference range 6-29 mg/l). Acetylcholinesterase isoenzyme gel electrophoresis showed a very strong B band, which was inhibited in the presence of BW 284C51. Ten days later an ultrasound B scan of the fetus showed a cystic structure 4 cm diameter on the posterior aspect of the skull and anteriorly a cyst compatible with encephalocele. The spine appeared intact with the exception of the cervical region, which was obscured by cyst. The fetal heart was noted to be beating. The patient was advised of the findings, and next day termination was performed by extra-amniotic prostaglandin E_2 .

The fetus was grossly hydropic, particularly over the occiput and nuchal region (figure). Over the occiput there was also a thin walled ruptured cyst, which did not, however, communicate with any intracranial structure at the base of the skull. Radiography showed no neural tube defect and no evidence of encephalocele. Further examination showed coarctation of the aorta and narrow "streak" ovaries. Chromosome analysis confirmed Turner's (45, XO) syndrome.

Comment

The maternal serum α fetoprotein concentration was not raised, though the concentration in the amniotic fluid was extremely high. This discrepancy is puzzling, but there are occasions when α fetoprotein concentrations may be pathologically high in amniotic fluid

Nineteen week old fetus with Turner's syndrome showing pronounced oedema in tissues of nuchal and cervical regions.

but not so in maternal serum owing to the widely differing protein sieving characteristics of amnions.¹

Ultrasound detected the hydropic "pterygium" over the neck and the small cyst over the occiput, which were interpreted as cystic structures compatible with encephalocele. A fluid filled sac over the occiput and nuchal area mistaken for a cervical meningocele was described by Sunderland in a full term infant with Turner's syndrome.² His studies of the sac fluid total protein and electrolytes showed this to be like a transudate of serum. Haddow *et al*³ reported the presence of an acetylcholinesterase band in studies of "hygromatous" fluid in a fetus with Turner's syndrome. Probably their reported "hygroma" was in fact oedematous tissue. Unfortunately, we did not study the oedema fluids from the "pterygium" in our case.

Raised amniotic fluid α fetoprotein concentration and an acetylcholinesterase band have been reported⁴ in pregnancy with Turner's syndrome of the fetus, though concentrations of the protein were not specified. The concentration of α fetoprotein in our amniotic fluid approached the expected concentration in fetal serum (roughly 150 times that of amniotic fluid in a normal pregnancy at an equivalent gestational age). Fetal serum (but not fetal red cells) gives a true positive test for acetylcholinesterase on gel electrophoresis.⁵ Fetal serum is the most likely source of acetylcholinesterase activity in the fluid filled oedematous tissues of fetuses with Turner's syndrome. The route by which the large amounts of α fetoprotein and excessive acetylcholinesterase activity reach amniotic fluid in Turner's syndrome, however, is still unknown.

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