Calcification of umbilical artery: two distinct lesions

TY KHONG,* S A DILLY

From the *Department of Paediatric Pathology, John Radcliffe Hospital, Oxford, and the Department of Histopathology, St George’s Hospital Medical School, London

SUMMARY The clinical and pathological features of five cases of calcification of umbilical cord vessels were reviewed. Two distinct lesions were identified: calcification could produce either sclerosis of the wall or obliteration of the lumen. In three cases there was calcification within the media and adventitia of the umbilical arteries, with extension into Wharton’s jelly in one case. The pathogenesis of this pattern of calcification—the sclerotic variant—is unclear but the findings of inflammation in the umbilical cord and its vessels, membranes, and decidua suggest intrauterine infection. In two cases there was complete calcification of umbilical arterial lumina resulting in total obliteration. The findings of fetal vessels in the chorionic plate with medial calcification in one of these two cases raises the possibility of thrombosis within the umbilical cord vessels as a cause, but the latter was not found. One infant from each group was liveborn. Both had shown signs of fetal distress in utero and delivered prematurely. The other three pregnancies resulted in macerated stillbirths preterm.

Calcification of umbilical cord vessels is rare. Previous reports have described clinical and pathological features associated with calcification of the vessel wall, but the paucity of reports suggest that it is an exceedingly rare occurrence. We report three further cases of umbilical vessel wall calcification and two cases of complete calcification of the umbilical arterial lumen, the latter phenomenon not previously reported.

Accepted for publication 20 April 1989

Table 1 Clinical data

<table>
<thead>
<tr>
<th>Case No</th>
<th>Age</th>
<th>Gravidity</th>
<th>Ethnic origin</th>
<th>Maternal history</th>
<th>Gestational age</th>
<th>Delivery</th>
<th>Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>24</td>
<td>G2P1</td>
<td>Occidental</td>
<td>Angioneurotic oedema, urticaria, polyarthritis lupus-like treated with prednisolone and azathioprine</td>
<td>35</td>
<td>Caesarean section for fetal distress</td>
<td>Liveborn male, 1480 g</td>
</tr>
<tr>
<td>2</td>
<td>19</td>
<td>G1</td>
<td>Occidental</td>
<td>Skin irritation and rash at 28 weeks gestation</td>
<td>34</td>
<td>Induced labour for intrauterine death</td>
<td>Macerated female, 940 g</td>
</tr>
<tr>
<td>3</td>
<td>43</td>
<td>G1</td>
<td>Oriental</td>
<td>Premature labour</td>
<td>32</td>
<td>Caesarean section for fetal distress</td>
<td>Liveborn male, 1350 g</td>
</tr>
<tr>
<td>4</td>
<td>22</td>
<td>G2P0 + 1</td>
<td>Occidental</td>
<td>—</td>
<td>25</td>
<td>Induced labour for intrauterine death</td>
<td>Macerated male, 619 g</td>
</tr>
<tr>
<td>5</td>
<td>21</td>
<td>G2P0 + 1</td>
<td>Occidental</td>
<td>—</td>
<td>32</td>
<td>Induced labour for intrauterine death</td>
<td>Macerated female, 1650 g</td>
</tr>
</tbody>
</table>
Table 2 Pathological findings

<table>
<thead>
<tr>
<th>Case No</th>
<th>Necropsy</th>
<th>Placenta</th>
<th>Histology</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Not relevant</td>
<td>292 g, 20% infarction</td>
<td>Complete calcification of umbilical arterial lumen</td>
</tr>
<tr>
<td>2</td>
<td>Permission refused; externally loss of fingers and toes due to amniotic rupture syndrome</td>
<td>460 g, cord twisted round partially detached amniotic epithelium</td>
<td>Thrombosis of fetal vessels in chorionic plate with medial calcification</td>
</tr>
<tr>
<td>3</td>
<td>Not relevant (umbilical artery catheter passed easily into preterm neonate)</td>
<td>393 g</td>
<td>Complete calcification of umbilical arterial lumen</td>
</tr>
<tr>
<td>4</td>
<td>Maceration changes</td>
<td>186 g, oedematous cord with narrowing at fetal insertion</td>
<td>Endarteritis with medial and adventitial proliferation and coarse calcification of one umbilical artery; fine calcification around the other two vessels</td>
</tr>
<tr>
<td>5</td>
<td>Maceration changes</td>
<td>360 g, umbilical cord showed marked circumferential fibrosis and calcification around all 3 vessels</td>
<td>Mild chorioamnionitis and acute deciduaitis present</td>
</tr>
</tbody>
</table>

Fig 1 Case 1: complete occlusion of arterial lumen by calcification. (Haematoxylin and eosin.)

Fig 2 Case 3: calcification of media and adventitia of umbilical arteries and vein. (Haematoxylin and eosin.)
of case 4. Calcification was seen in Wharton’s jelly in case 5 (fig 3). Thrombosis of fetal vessels in the chorionic plate with either calcification of the media or the contents was seen in cases 2 and 5 (fig 4). Gram stains did not show the presence of organisms.

**Discussion**

Because placentas are examined on a semi-selective basis the exact prevalence of calcification of umbilical cord vessels is not known. We believe, however, that the condition is rare as these five cases were found from archival material over a 13 year period (1975–1988) in Oxford and over a five year period (1983–1988) at St George’s Hospital, London.

Calcification of umbilical cord vessels can take one of two forms producing obliteration on the one hand and sclerosis on the other. deSa distinguished between calcification of umbilical cord vessels and sclerosing funisitis or constrictive sclerosis of Wharton’s jelly. In the former, there is calcification of the cord vessel wall while in the latter there is often calcification of an attenuated and sclerosed Wharton’s jelly. Our third and fourth cases showed calcification predominantly in the adventitia while our fifth case showed calcification within Wharton’s jelly as well as within the wall of the blood vessels; it is our contention that it is difficult to ascertain the primary site of calcification. This is not surprising as the adventitia merges imperceptibly with the surrounding Wharton’s jelly and Wharton’s jelly has been likened to the adventitia of the umbilical arteries. It would seem more appropriate to classify these as sclerosing calcification in contrast to the obliterative lesion seen in cases 1 and 2. Blanc suggested that calcified umbilical cord vessels were due to intrauterine viral infections while deSa thought that sclerosing funisitis was a response to any infection. In none of our cases, however, was there evidence of villitis or viral inclusions, but the findings of decidualitis (case 3), funisitis (cases 4 and 5), chorioamnionitis (cases 3 and 4) and endarteritis or vasculitis (cases 3, 4, and 5) suggest intrauterine infection. Bacteriological studies on the mothers were negative. Sclerosing calcification of umbilical vessels has been found in association with fetal hydrops following fetomaterial haemorrhage, Salla disease, fetal renal calcification, normal liveborn infants and macerated stillbirths.
Complete calcification of the umbilical artery lumen as seen in our first two cases has not hitherto been described. Its pathogenesis is unclear but the finding of focal calcification of thrombus of the major vessels in the chorionic plate in case 2, similar to those described by Benirschke and Driscoll, suggests that it may be a degenerative phenomenon, as a result of dystrophic calcification of a thrombus more proximally. Perrin and Kahn-Vander Bel presented a case in which there was focal calcification of a laminated thrombus in an umbilical cord vessel while Heifetz had a case of “vascular mural calcification” in his series of umbilical cord thrombosis, although the extent of calcification was not illustrated. Thrombosis of umbilical vessels is rare, and a thrombus was not found in the umbilical cords of cases 1 or 2.

These cases are different to the condition referred to as idiopathic arterial calcification or infantile arterial calcification where generalised arterial calcification, including coronary and renal arteries, with or without intimal proliferation, have been described. In that condition, some were stillbirths but most were infants and in none has calcification of the umbilical arteries been described. Furthermore, in our cases 4 and 5 where necropsy was performed, systemic calcification was not seen.

Whatever the pathogenesis of calcification of umbilical cord vessels, there seem to be two relatively distinctive patterns, either calcification leading to complete obliteration of the arterial lumen or calcification of the arterial wall with or without involvement of Wharton’s jelly. A possible cause for the first is thrombosis and, for the second, infection.

We thank the clinicians Mr D H Barlow, J M Pearce, J H Hughes, S C Simmons for their permission to report these cases, Dr J W Keeling for helpful discussions, and Mr G Richardson and Miss H Mellor for photographic assistance.

References


Requests for reprints to: Dr T Y Khong, Department of Paediatric Pathology, John Radcliffe Hospital, Oxford OX3 9DU, England.
Calcification of umbilical artery: two distinct lesions.

T Y Khong and S A Dilly

doi: 10.1136/jcp.42.9.931

Updated information and services can be found at:
http://jcp.bmj.com/content/42/9/931

These include:

**Email alerting service**

Receive free email alerts when new articles cite this article. Sign up in the box at the top right corner of the online article.

Notes

To request permissions go to:
http://group.bmj.com/group/rights-licensing/permissions

To order reprints go to:
http://journals.bmj.com/cgi/reprintform

To subscribe to BMJ go to:
http://group.bmj.com/subscribe/