PULMONARY EMBOLISM IN CHILDREN

BY

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Pulmonary embolism is generally believed to be rare in childhood, and my own survey of the published material suggests that under 50 cases have so far been recorded. Stevenson and Stevenson (1949), who surveyed published reports from all over the world and reported a case in 1949, could find only 30 reports of which only 16 had had complete autopsies. Not all cases, however, are fatal. Cashman (1956) reported an embolus in a boy of 9 years who recovered completely.

The first reported case appears to have been almost exactly a century ago when Löschner (1861) described a pulmonary embolus in a 9-year-old boy, which followed phlebitis associated with a fracture of the tibia. His case typifies many of the earlier case reports of the pre-antibiotic days where emboli appeared to follow injury to bones and muscles (Rosenbloom and Henderson, 1935). Rupp (1921) tabulated 11 cases of fatal pulmonary embolism that had followed prolonged wasting disease in children under the age of 10 years. Several cases have been reported following prolonged or acute sepsis (Wessén, 1922; Fleischmann, 1929; Stulik and Rust, 1929; Jones, 1930; Hosoi, 1932).

My own interest in pulmonary emboli in children was stimulated by the symptoms and lethal complication of the ventricular-atrial Spitz Holter valve in children with hydrocephalus (Emery and Hilton, 1961). Many of these children succumb from pulmonary embolism from clots that arise around the tube in the right atrium. All cases of emboli associated with the Spitz Holter valve or with primary disease of the heart have, however, been excluded in the present survey.

This report concerns 25 cases of fatal pulmonary embolism that we have seen in our own material during the past 10 years, and an attempt is made to assess its importance in the pathology of the lung in children.

Material and Methods

The cases surveyed here came from a series of approximately 2,000 autopsies from liveborn children that have been carried out by the Department of Pathology at the Sheffield Children’s Hospital. These child deaths comprise not only hospital admissions but also many deaths at home, referred into hospital by practitioners for autopsy, and children dying in such circumstances as have been referred to the Coroner. The routine practice at autopsy in this hospital has been designed to fix as much of the tissues of the body as possible before any manipulation has taken place. The technique concerning heart and lungs has been to open the superior and inferior vena cava while the heart and lungs are within the chest to ensure that the systemic venous return has been normal and there are no gross deformities in the vascular return to the heart. After removing the tongue, heart and lungs, an incision is made through into the right ventricle. The ventricular septum is carefully inspected and the pulmonary valve opened, including a short length of the main pulmonary trunk, but the blood throughout the pulmonary arterial system is not removed and the heart and lungs from this point onwards are placed in formalin for a period of approximately 10 days before further dissection.

After fixation, blocks are taken from each lobe of the lung, the blocks being transverse to the general line of the main bronchi and vessels. Sections are routinely stained with Masson’s trichrome.

In all but three cases (Fig. 1), the diagnosis of pulmonary embolism has rested upon the histological diagnosis of ante-mortem thrombi in arteries, the thrombi being of such form and structure that they could not have arisen as local thrombi (Fig. 2).

The histological diagnosis of ante-mortem thrombosis in situ is not easy, particularly if the thrombosis is of recent origin. I have excluded some equivocal cases. For the diagnosis of an ante-mortem thrombosis there must be streaming and lamination of fibrin; the state of blood in other vessels is used in comparison. The diagnosis of emboli in the pulmonary arteries is easier and more certain.

Results

Some details of the 25 cases are set out in the Table.

In these children the history falls into two definite phases: first, the prodromal symptoms and disease, i.e. the disease associated with the onset and cause of the primary thrombosis; and secondly, the symptoms apparently associated with the incident
of the embolism and those intervening between that time and death. Since the present series only concerns morbid anatomy, possible cases of embolism with recovery have not been discussed or included, although such cases have been reported and are probably much more common than is usually appreciated.

The cases fall into two age-groups. There is a neonatal group of five children. Two of these deaths followed cerebral venous thrombosis and two were secondary to thrombi in the venous channels of the left liver. The following case report is an example of this.

Case 3. This was a full-term child born in blue cyanosis. She improved in colour during the next few hours, but later on the day of birth she became shocked and remained apparently in this shocked state for approximately 24 hours. She was admitted to hospital in extreme cyanosis and died almost immediately, three days after birth. Autopsy showed deep red lungs but not heavy as in hyaline membrane disease. Histology revealed emboli in many of the pulmonary arteries. The left lobe of the liver was deep red and in a state of partial collapse with many thrombosed hepatic veins, the thrombi being very similar to the emboli seen in the lungs. In this case the primary cause of the embolism was apparently the shocked state of the child, which made it unable to maintain an adequate circulation through the left liver.
<table>
<thead>
<tr>
<th>Case No.</th>
<th>Age at Death</th>
<th>Prodromal Disease</th>
<th>Terminal Symptoms and Duration</th>
<th>Primary Site of Thrombosis</th>
<th>Naked-eye Emboli in Pulmonary Trunk</th>
<th>Emboli in Pulmonary Arteries</th>
<th>Thrombi in Pulmonary Veins</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Days 2</td>
<td>Exomphalos; bowel resection</td>
<td>Distressed breathing, blood in stomach, flaccid; 1 hour</td>
<td>Renal vein and I.V.C. and mesentery</td>
<td>-</td>
<td>+</td>
<td>-</td>
</tr>
<tr>
<td>2</td>
<td>3 days</td>
<td>‘Mucousy’; tracheoesophageal fistula</td>
<td>One day after operation; sudden death</td>
<td>Left liver</td>
<td>-</td>
<td>+</td>
<td>-</td>
</tr>
<tr>
<td>3</td>
<td>3 months</td>
<td>Shocked</td>
<td>Sudden severe cyanosis; 4 hours</td>
<td>Left liver</td>
<td>-</td>
<td>+</td>
<td>-</td>
</tr>
<tr>
<td>4</td>
<td>6 months</td>
<td>Cerebral irritation</td>
<td>Convulsions for 12 hours</td>
<td>Cerebral sinus</td>
<td>-</td>
<td>+</td>
<td>+</td>
</tr>
<tr>
<td>5</td>
<td>7 weeks</td>
<td>Jaundice; cerebral irritation</td>
<td>Found dead in cot in hospital 2 days after admission</td>
<td>Cerebral</td>
<td>-</td>
<td>+</td>
<td>-</td>
</tr>
<tr>
<td>6</td>
<td>2 months</td>
<td>Skin rash and sniffles</td>
<td>Bleeding from nose, sudden death ½ hr later</td>
<td>Inferior vena cava and renal</td>
<td>-</td>
<td>+</td>
<td>+</td>
</tr>
<tr>
<td>7</td>
<td>2 weeks</td>
<td>Sticky eye, 'unwell'</td>
<td>Found dead</td>
<td>Nasopharyngeal</td>
<td>-</td>
<td>+</td>
<td>+</td>
</tr>
<tr>
<td>8</td>
<td>3 weeks</td>
<td>Nasal discharge</td>
<td>Sudden death</td>
<td>Cerebral sinus</td>
<td>-</td>
<td>+</td>
<td>-</td>
</tr>
<tr>
<td>9</td>
<td>3 weeks</td>
<td>Off food Acute abdominal pain</td>
<td>Sudden, unexpected death within an hour of laparotomy</td>
<td>Pelvis (sepsis)</td>
<td>-</td>
<td>+</td>
<td>+</td>
</tr>
<tr>
<td>10</td>
<td>4 weeks</td>
<td>Off food Colic and diarrhoea</td>
<td>Sudden death in mother's arms after period of extreme restlessness</td>
<td>Cerebral</td>
<td>-</td>
<td>+</td>
<td>-</td>
</tr>
<tr>
<td>11</td>
<td>4 weeks</td>
<td>Off food</td>
<td>Found dead at home in cot</td>
<td>Not found</td>
<td>-</td>
<td>+</td>
<td>-</td>
</tr>
<tr>
<td>12</td>
<td>5 weeks</td>
<td>Skin sepsis ‘Purpura necrotica’</td>
<td>Sudden extreme dyspnoea for 1 hour</td>
<td>Cerebral and renal</td>
<td>-</td>
<td>+</td>
<td>-</td>
</tr>
<tr>
<td>13</td>
<td>6 weeks</td>
<td>Nasal discharge</td>
<td>Sudden severe dyspnoea lasting 4 hours</td>
<td>Not found</td>
<td>-</td>
<td>+</td>
<td>?</td>
</tr>
<tr>
<td>14</td>
<td>7 weeks</td>
<td>Cough Vomiting Convulsions</td>
<td>Sudden death in hospital 1 day after admission</td>
<td>Cerebral</td>
<td>-</td>
<td>+</td>
<td>+</td>
</tr>
<tr>
<td>15</td>
<td>8 weeks</td>
<td>Sore throat</td>
<td>Acute respiratory distress for 8 hours</td>
<td>Nasopharyngeal</td>
<td>-</td>
<td>+</td>
<td>+</td>
</tr>
<tr>
<td>16</td>
<td>9 weeks</td>
<td>Cough</td>
<td>Acute collapse and cyanosis; sudden death 2 days after admission</td>
<td>Cerebral</td>
<td>-</td>
<td>+</td>
<td>+</td>
</tr>
<tr>
<td>17</td>
<td>10 weeks</td>
<td>Refusing food</td>
<td>Convulsions for 6 hours</td>
<td>Cerebral</td>
<td>-</td>
<td>+</td>
<td>-</td>
</tr>
<tr>
<td>18</td>
<td>11 weeks</td>
<td>Abdominal pain — intestinal obstruction</td>
<td>Sudden death in hospital 6 weeks following operation</td>
<td>Pelvic veins and vena cava</td>
<td>+</td>
<td>+</td>
<td>-</td>
</tr>
<tr>
<td>19</td>
<td>16 weeks</td>
<td>Cold</td>
<td>Severe respiratory distress, asthma for 4 hours</td>
<td>Not known</td>
<td>-</td>
<td>+</td>
<td>+</td>
</tr>
<tr>
<td>20</td>
<td>23 weeks</td>
<td>Acrodynia; ulceration of gums</td>
<td>Found dead in bed having been in hospital several weeks</td>
<td>Inferior vena cava and renal</td>
<td>+</td>
<td>+</td>
<td>-</td>
</tr>
<tr>
<td>21</td>
<td>Years 2</td>
<td>Vomiting</td>
<td>Acute respiratory distress for 12 hours</td>
<td>Cerebral and renal</td>
<td>+</td>
<td>+</td>
<td>?</td>
</tr>
<tr>
<td>22</td>
<td>2 weeks</td>
<td>Poliomyelitis injection in buttocks Tonsillitis</td>
<td>Pain in thigh; sudden death in parent's arms</td>
<td>Deep gluteal pelvis</td>
<td>+</td>
<td>+</td>
<td>-</td>
</tr>
<tr>
<td>23</td>
<td>3 months</td>
<td>Adrenogenital syndrome Vomiting and tonsillitis</td>
<td>Sudden collapse and death 2 days after admission to hospital</td>
<td>Nasopharyngeal</td>
<td>-</td>
<td>+</td>
<td>+</td>
</tr>
<tr>
<td>24</td>
<td>3 weeks</td>
<td>Vomiting and nasal discharge</td>
<td>Sudden death in parent's arms</td>
<td>Cavernous sinuses and nasopharyngeal</td>
<td>-</td>
<td>+</td>
<td>+</td>
</tr>
<tr>
<td>25</td>
<td>7 months</td>
<td>Nasopharyngeal tumour Nasal sepsis</td>
<td>Sudden death having been in hospital many weeks</td>
<td>Nasopharynx</td>
<td>+</td>
<td>+</td>
<td>-</td>
</tr>
</tbody>
</table>
Children of older age varied from 13 weeks to 7 years.

The primary sites of venous thrombosis appear to be the cerebral venous sinuses in nine cases, nasopharyngeal in five, lower abdominal and pelvic in four, and inferior vena cava and renal in two. In three instances the site of primary thrombosis was not discovered.

Only three of the cases were post-operative, one in a 3-day-old child who collapsed and died suddenly some six hours after operation for repair of a tracheo-oesophageal fistula; the site of thrombosis was the left liver. Two other cases occurred after abdominal sepsis and were associated with pelvic and retroperitoneal abscesses, one was secondary to an intestinal perforation and the other associated with a urachal cyst and abscess. The small numbers of post-operative deaths in this series are probably significant in view of the fact that more than half of the beds in the hospital were for acute surgery, and there was a very rapid turnover.

Of the older children, less than one-half presented as sudden and unexpected deaths. The others chiefly presented as very acute infections of the lung, and the following is a report of one such child.

Case 19. This was a 16-month-old child who was admitted as an emergency for what was thought to be an attack of acute asthma. There was no response to adrenaline, and the child died within two hours of admission. Although ante-mortem emboli were found in the pulmonary arteries in this child, the primary site of thrombosis was not identified.

A particular group was related to infection of the nasopharynx. Such was a boy of a little over 3 years (Case 23).

Case 23. He was admitted because of vomiting. He had shown adrenogenital syndrome as a newborn and had been well maintained on corticosteroids. He had grossly enlarged tonsils and severe inflammation of the pharynx. Two days after admission, he collapsed and died suddenly. Autopsy showed the lungs to be congested, and the only other abnormality was an acute inflammation of the tonsils and nasopharyngeal tissue. Histology showed venous thrombosis in veins in the peritonsillar tissues, emboli in the pulmonary arteries and some thrombi in pulmonary veins. There was little doubt that the immediate cause of death in this child was pulmonary embolism from clots formed in the vessels of the nasopharynx.

Case 24. This boy, who had achondroplasia and was 33 years of age, one day vomited several times. He was seen by his doctor and found to have pyrexia with pus coming from his nose. The practitioner said that the boy had a severe pharyngitis and tonsillitis and gave him oral penicillin. In the early afternoon of the following day he had a deep sleep and when he awoke he again vomited several times. Later that evening his father picked the boy up in his arms to take him to bed, and he suddenly kicked, fought for breath and died in his father's arms. At autopsy the only abnormal features were congestion of the lungs and extreme oedema of the perinasal tissues with possible early quinsy, but there was no obstruction of the airways. Later $\beta$-haemolytic streptococci were grown from the nasal cavities. An ante-mortem thrombosis of the cavernous sinus was confirmed histologically and a large twisted embolus was found packed in the main pulmonary artery (Fig. 2). Thrombosed vessels were also found in the peritonsillar tissue and also associated with some of the deep cervical lymph nodes.

A further group of cases was associated with cerebral symptoms, some of these related to trauma.

Case 8. A 3-month-old child who had apparently been quite fit had fallen from his cot onto the floor on the day before death. The following morning he took three-quarters only of his feed and after bringing up wind, suddenly became cyanosed, limp and died in his mother's arms. Autopsy revealed the presence of a recent superficial but extensive subdural haematoma up to 5 mm. in thickness. The haemorrhage had come from a torn vein running into the longitudinal sinus. There were small blood clots within the venous cerebral system. No clot was found within the longitudinal sinus itself, but only in the vessels running into it. The lungs showed the presence of emboli in the pulmonary arteries and a few ante-mortem clots within the pulmonary veins. This child had apparently died from pulmonary embolism and thrombosis secondary to clots probably coming from the longitudinal sinus.

Discussion

Venous thrombosis is by no means an uncommon finding at autopsy in children, and, in a series of 1,000 autopsies that we recently surveyed, 81 were found in whom thrombosis of veins in some part of the body was considered to be a factor contributing to death. Thrombosis in pulmonary vessels is also not rare particularly associated with symptoms usually considered to be those of a very severe respiratory infection (Emery, 1953) and probably is as common in children as in adults (Spain and Moses, 1946). It is these thrombi that are probably the major factor in the lethal outcome of a chest infection. The role of embolism in such cases is difficult to elucidate. Within our own material of known cases of embolism, we have found that if severe respiratory symptoms have existed for more than a matter of two to three hours, a pulmonary embolus is usually associated with pulmonary venous thrombosis. In general, the only cases in which we have not found pulmonary venous
thrombosis associated with pulmonary embolism, are those that have died apparently from the shock of the embolism.

The symptoms following pulmonary embolism are very like those of an acute infection of the respiratory tract (Emery and Hilton, 1961). This was a feature of the child deaths following embolism from clots around the auricular end of the tube in the Spitz Holter valve shunt and largely stimulated the present survey. In those children the diagnosis in the early cases was that of 'acute pneumonia', and we suspect that many of the deaths reported from such children as being due to acute bronchopneumonia were in fact embolism deaths.

The reason why we have found so many cases of pulmonary embolism is probably due to the technique used at autopsy. On simple gross dissection we have only found three cases; the other 21 cases came from microscopy of the pulmonary vessels, the vessels not having been separated from the heart and thus emptied of their blood before histological examination. It is possible that, if more detailed and thorough examination had been carried out, even greater numbers of pulmonary emboli would have been discovered.

Pulmonary embolism is never a primary disease but an accidental complication of other diseases in children where there is venous thrombosis (Zuelzer, Kurnetz and Fallon, 1951). In our own material common precipitating lesions have been cerebral haemorrhage or cerebral thrombosis, sepsis in the lower abdomen, cachexia or acute severe nasopharyngeal infection. Only approximately one-third of the cases that have come into our hands have died in a sudden dramatic way; the others have had an abrupt onset of severe respiratory symptoms. It seems likely that a number of cases of pulmonary embolism in children do survive, such as the case reported by Cashman (1956), but many are undiagnosed or are diagnosed as acute pneumonia. It is not the function of this paper to discuss therapy, but to draw attention to the possibility of pulmonary embolism in children. Whenever unexpected symptoms of infection of the lung arise in a child, under treatment for disease in some other part of the body, pulmonary embolism and thrombosis should be considered.

Summary

The clinical symptoms of pulmonary embolism in children are usually those of an acute infection of the respiratory tract. Common sites of origin of emboli in babies under 1 month of age are hepatic veins and, in older children, the cerebral sinuses and the veins of the nasopharynx.

References

Pulmonary Embolism in Children

John L. Emery

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